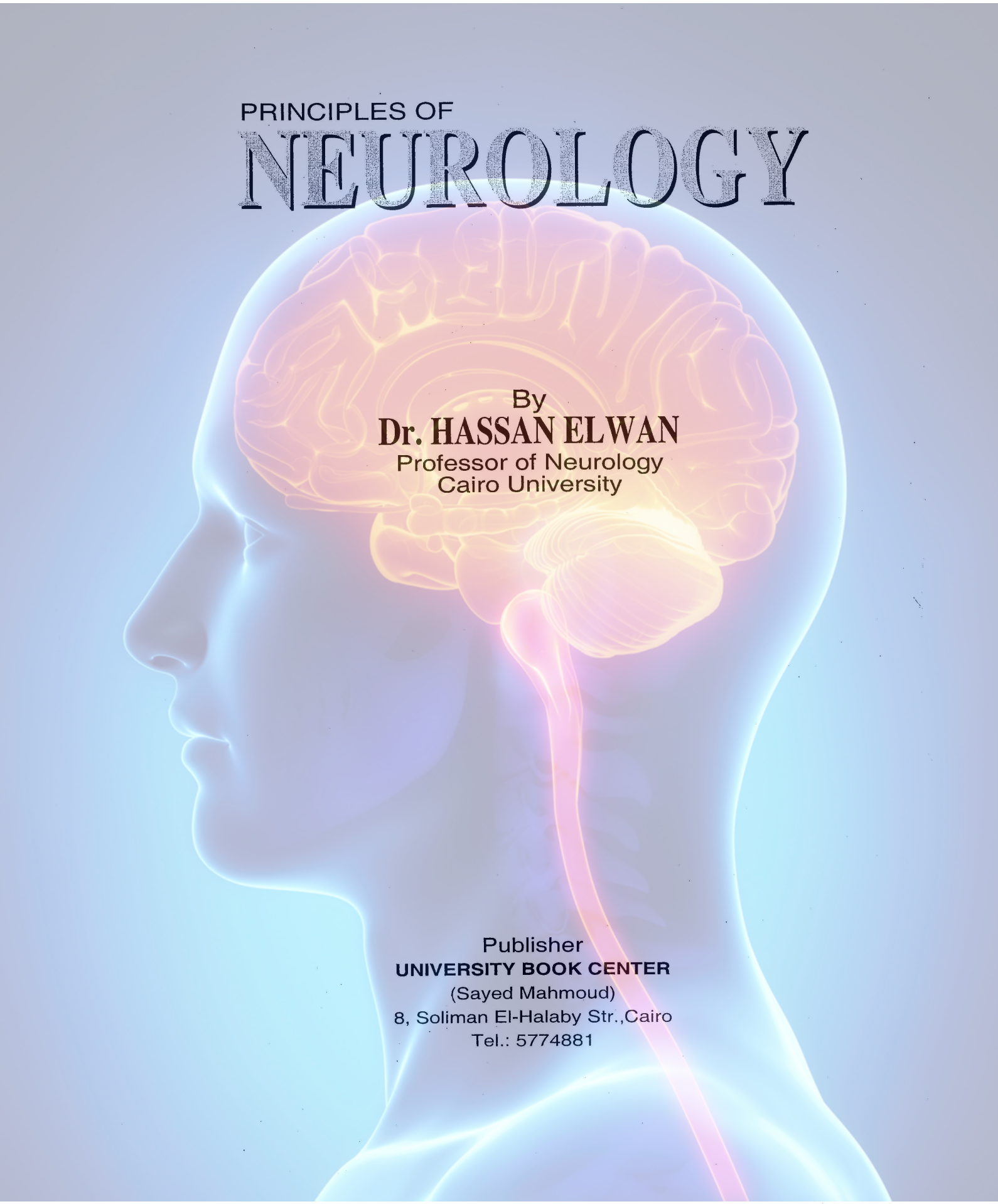
PRINCIPLES OF

NEUROLOGY



By

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INTRODUCTION

The Central Nervous System (C.N.S.) is formed of 2 main parts:

1) INTRACRANIAL PART formed of:

A- Cerebrum B-Brain Stem C- Cerebellum.

A. Cerebrum:

- It is formed of **2 cerebral hemispheres**, connected to each other by the corpus callosum, and to the upper part of the brain stem by the 2 cerebral peduncles.
- The surface of each hemisphere is divided into 4 lobes:
 1. Frontal 2. Parietal 3. Temporal 4. Occipital.
- The lobes are separated from each other by **sulci** (fissures):
 The Central Sulcus separates the frontal from the parietal lobe.
 The Parieto-occipital Sulcus separates the parietal from the occipital lobe.
 The Lateral Sulcus (Sylvian fissure) separates the frontal & parietal lobes from the temporal lobe.
- The cerebral lobes are formed of:
 1. Outer **gray matter** composed of nerve cells (**cerebral cortex**). It contains certain **areas,** the cells of which are concerned with specific functions e.g. the Motor area (area 4) is concerned with the initiation of voluntary motor activity (see next chap.).
 2. Inner **white matter** composed of nerve fibres conducting impulses to & from the cells of the cortex.
- At the base of each cerebral hemisphere, there are several groups of nuclei situated at various levels within the white matter; they form the basal ganglia, thalamus, subthalamus and hypothalamus.

B. Brain Stem:

- It is formed of: 1. **Midbrain** **2. Pons** **3. Medulla.**
- It is connected to the cerebral hemispheres by 2 cerebral peduncles and to the cerebellum, on each side, by the superior, middle & inferior cerebellar peduncles.
- It contains groups of nerve cells (gray matter) intermingled with several ascending and descending fibres (white matter).
- The **Motor nuclei of the Cranial Nerves** are arranged in the Brain Stem as follows:

Cr 3 & 4	in Midbrain
Cr 5, 6 & 7	in Pons
Cr 9, 10, 11 & 12	in Medulla.

N.B. Cr 1, 2 & 8 are sensory nerves concerned with special sensations, perceived in special areas of the cerebral cortex.
Cr 1,2 & 8 have no motor nuclei.

C. Cerebellum:

It lies behind the Brain Stem and occupies most of the posterior cranial fossa. It is concerned with coordination of voluntary motor activity and maintenance of equilibrium.

2) SPINAL PART formed of:

A- Spinal Cord **B- Cauda Equina.**

A. Spinal Cord:

- It lies in the spinal canal & ends at the lower border of the 1st lumbar vertebra.
- The lowermost 3 segments of the spinal cord (S3, 4, 5) are known anatomically as the **conus medullaris** while the above 4 segments (L4, 5, S1, 2) are known anatomically as the **epiconus**.
- It is formed of **gray matter** (cells) surrounded by **white matter**.
- In a transverse section the gray matter resembles the letter H (2 anterior & 2 posterior horns).
- The white matter contains ascending and descending nerve fibres arranged into **tracts**. The important tracts are:

ASCENDING TRACTS	DESCENDING TRACTS	
1. Lateral spinothalamic	1. Corticospinal (pyramidal)	
2. Ventral spinothalamic	2. Reticulospinal	Extra-pyramidal
3. Gracile & Cuneate	3. Vestibulospinal	
4. Spinocerebellar	4. Tectospinal	
5. Lissauer's tract.	5. Rubrospinal	
	6. Olivospinal	

B. Cauda Equina:

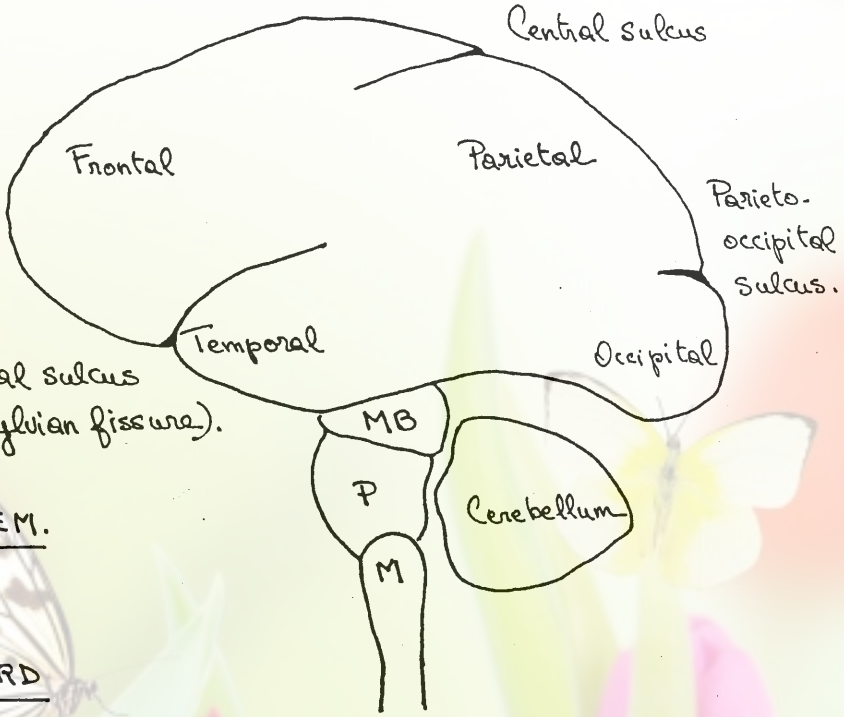
It is the collection of lumbosacral roots which fill the lower part of the spinal canal below the lower border of L1 vertebra.

PARTS OF CNS

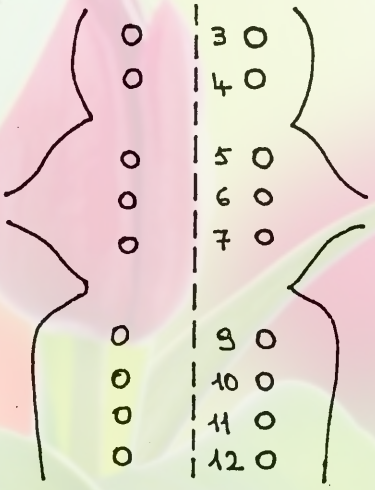
I. CEREBRUM.
(Cerebral cortex)

II. BRAIN STEM.

III. SPINAL CORD



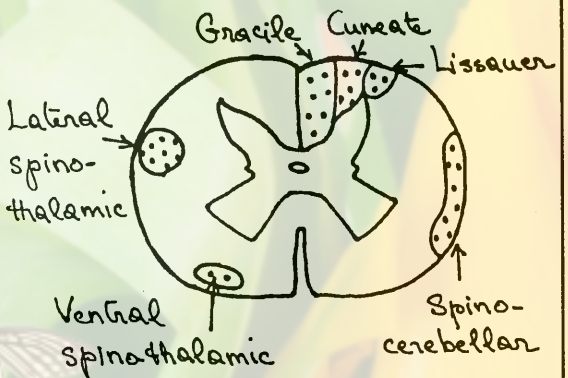
BRAIN STEM



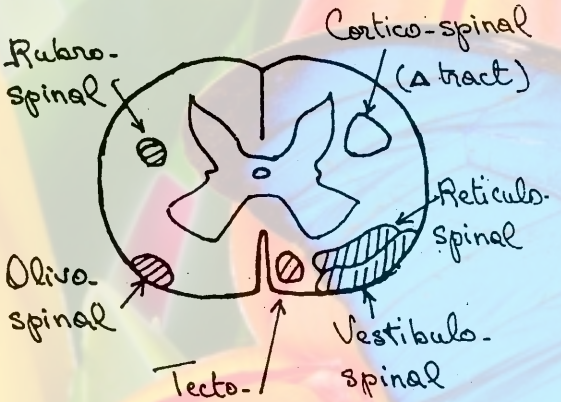
Motor Nuclei of cranial nerves

* Cranial 1, 2, 8 are only sensory.

SPINAL CORD



~~Ascending Tracts.~~



~~Descending Tracts.~~

CEREBRAL CORTEX

A. FRONTAL LOBE

1) Motor Area (area 4):

- Site: floor of central sulcus & posterior part of precentral gyrus.
- Function: initiation of voluntary motor activity of the opposite $^1/_2$ of the body through the pyramidal (Δ) tract. **In this area the body is represented upside down.** Complex movements involving speech, face & hands are widely represented in the lower part of this area.
- Lesion: – Irritative: contralateral **motor Jacksonian fits**: there are convulsions involving the muscles of one side of the body; the fit has a focal onset either in the thumb, angle of the mouth or big toe (depending on whether the irritative lesion starts in the lower or upper part of the motor area); the fit spreads in a march course e.g. thumb → arm → shoulder → trunk → L.L.
 – Destructive: Contralateral **paralysis** usually affecting one limb (monoplegia).

2) Premotor Area (area 6):

- Site: anterior part of the precentral gyrus.
- Function: 1- partly supplies Δ tract & 2- gives extra Δ fibres.
 This area inhibits the muscle tone & the deep reflexes on the opposite side of the body.
- Lesion: 1- contralateral **hypertonia & exaggerated deep reflexes**.
 2- contralateral **fanning** of the lateral 4 toes on eliciting the Plantar reflex.

3) Area of Voluntary Conjugate Eye Movements (area 8):

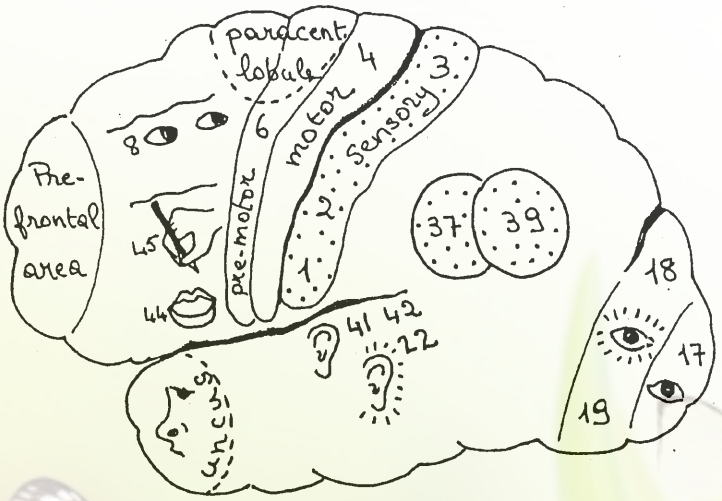
- Site: posterior part of middle frontal gyrus.
- Function: voluntary conjugate eye movement to the opposite side e.g. while reading the action of passing from the end of one line to the beginning of the next line; this movement is usually rapid & is termed "**saccadic**."
- Lesion: – Irritative: attacks of conjugate eye deviation to the opposite side of the lesion.
 – Destructive: paralysis of conjugate eye movement to the opposite side of the lesion.

4) Broca's Area (area 44):

- Site: posterior part of inferior frontal gyrus of **DOMINANT** hemisphere.
- Function: motor centre for speech.
- Lesion: **motor** (expressive) **aphasia**; the patient cannot express his ideas in spoken words. (see p 37).

5) Exner's Area (area 45):

- Site: adjacent to area 44 in the **DOMINANT** hemisphere.
- Function: centre for writing.
- Lesion: **agraphia**; the patient cannot express his ideas in written words.



AREAS OF THE CERBERAL CORTEX

6) Pre-Frontal Area:

- Site: anterior part of frontal lobe & its adjacent inferior surface.
- Function: 1- higher centre for mentality, personality & behaviour.
 2- inhibition of primitive reflexes which are present in the newborn. e.g. grasp, grope, pout, glabellar & palmo-mental reflexes (see p. 146).
- Lesion: 1- mentality, personality & behavioural changes: lack of attention & judgement, disinterest in people & surroundings, lack of personal hygiene, ending in dementia.
 2- reappearance of primitive reflexes.

7) Paracentral Lobule:

- Site: medial surface of the superior frontal gyrus, adjacent to the foot & leg area.
- Function: cortical inhibition (control) of bladder & bowel voiding
- Lesion: incontinence of urine & faeces.

B. PARIETAL LOBE

1) Cortical Sensory Area (areas 1, 2, 3):

- Site: post-central gyrus.
- Function: Perception of cortical sensations from the opposite $^1/_2$ of the body; like in the motor area, the body is represented upside down.
- Lesion: – Irritative: contralateral **sensory Jacksonian fits** in the form of numbness or tingling with focal onset & a march course; it may be followed by a motor fit if the irritation extends to the adjacent motor area.
 – Destructive: contralateral **cortical sensory loss**.

2) Angular Gyrus (area 39):

- Site: in the postero-inferior part of the parietal lobe.
- Function: in the **DOMINANT** hemisphere, it is concerned with reading i.e. the recognition & recall of letters & numbers.
- Lesion: **Alexia**; the patient who could read before the lesion, becomes unable to do so, because he cannot understand the letters & numbers which he sees.

3) Supramarginal Gyrus (area 37):

- Site: anterior to the angular gyrus.
- Function: in the dominant hemisphere it is concerned with storage & recall of:
 - IDEAS of speech.
 - IDEAS of complex voluntary motor activity.
- Lesion: 1- **Jargon's aphasia** (word salad).
 2- **Apraxia**: inability to perform complex voluntary motor activity in absence of paralysis, incoordination or sensory loss.

C. TEMPORAL LOBE

1) Auditory Sensory Area (area 41, 42):

- Site: superior temporal gyrus.
- Function: auditory sensory area.
- Lesion:
 - Irritative: auditory hallucinations.
 - Destructive: Slight hearing impairment, **never deafness** as hearing is bilaterally represented.

2) Auditory Associative Area (area 22):

- Site: adjacent to areas 41 & 42.
- Function: recognition & recall of sounds.
- Lesion: **Auditory agnosia**: the patient hears but does not understand (recognize) what he hears.

3) Limbic System:

- Site: uncus & hippocampus in the medial & inferior surfaces of the temporal lobe.
- Function: concerned with smell (uncus), mood & memory.
- Lesion: 1. **Uncinate fits** with olfactory hallucinations, usually unpleasant.
 2. **Temporal lobe seizures** (see epilepsy).
 3. **Anterograde amnesia** (loss of memory for recent events).

D. OCCIPITAL LOBE

1) Visual Sensory Area (area 17): for the reception of visual images.

2) Visual Associative Area (area 18, 19): anterior to area 17.

- Function: 1- recognition & recall of images.
 2- centre for reflex conjugate eye movement to the opposite side **e.g.** while reading, following the words of a line, one after the other; this movement is usually slow & is termed "**pursuit**."
- Lesion to the visual areas results in:
 - Irritative: **Unformed visual hallucinations** e.g. sparks, lines, flashes ...; this occurs e.g. in the aura of classic migraine or in epilepsy when the occipital lobe is involved.
 - Destructive: 1- **Homonymous hemianopia** with or without macular sparing.
 2- **Visual Agnosia**: the patient sees (e.g. a familiar face) but does not recognize what he sees.
 3- **Paralysis of reflex conjugate eye movements**.

THE MOTOR SYSTEM

The motor system consists of 4 main components:

1. The Pyramidal (Δ) System (U.M.N.):

It originates in the motor area (4) & premotor area (6) & terminates at the anterior horn cells (A.H.C.) of the different levels of the spinal cord. It supplies the opposite side of the body.

2. The Extrapyramidal (extra Δ) System:

It originates from centres situated at various levels of C.N.S. mainly the Basal Ganglia.

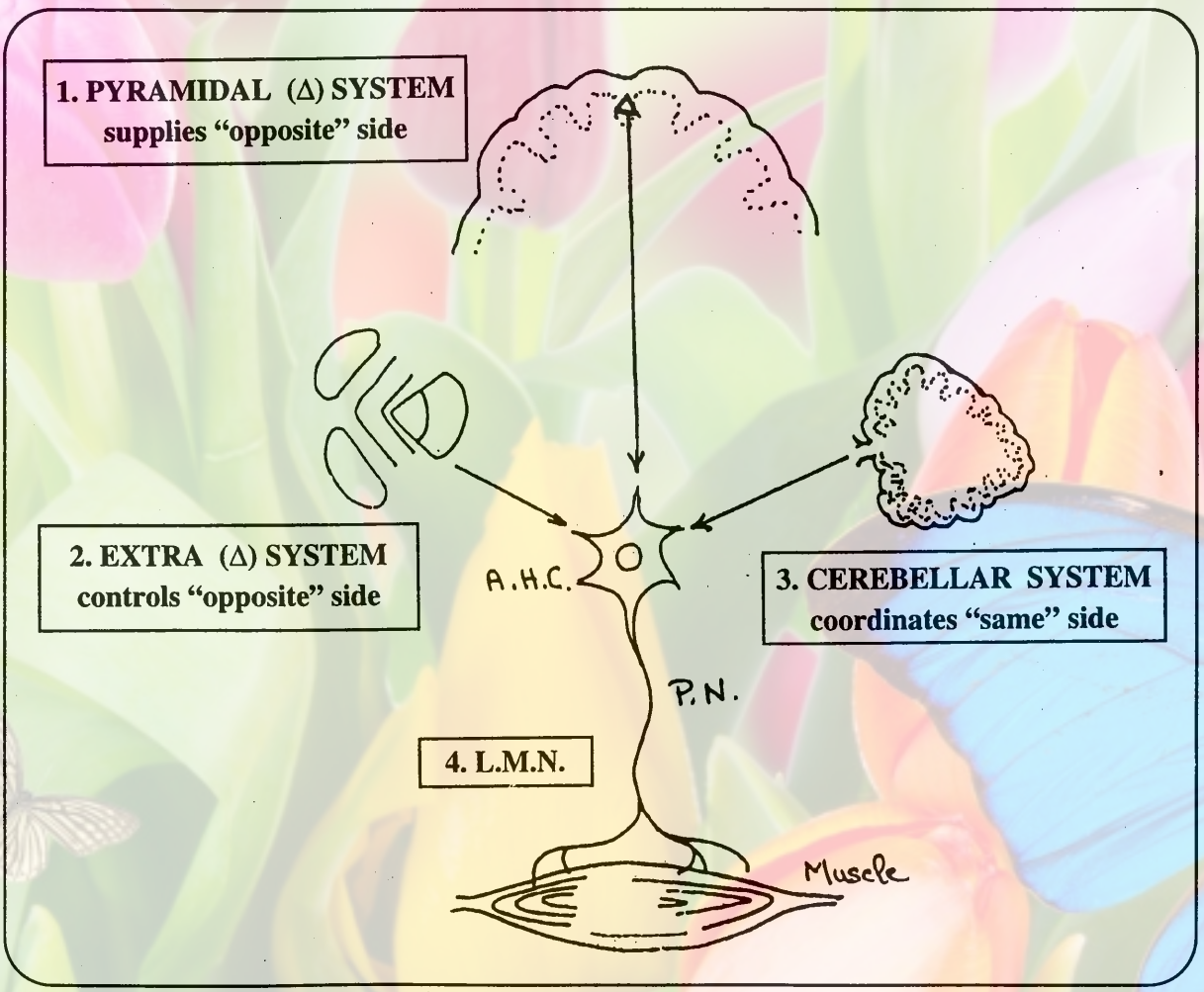
It controls the opposite side of the body.

3. The Cerebellar System:

It is composed of the Neo, Archi & Paleo-cerebellum. It coordinates the movements of the same side of the body.

4. The Lower Motor Neurone (L.M.N.)

It is formed of A.H.C.s & peripheral motor nerves (which transmit the motor impulses to the voluntary muscles).



UPPER & LOWER MOTOR NEURONES

A neurone is formed of:

1. Nerve cell.
2. its Axon that ends
 - around another nerve cell (synapse) or
 - at the motor end plate of a muscle.

PATHWAY OF VOLUNTARY MOTOR IMPULSE:

For a voluntary muscle to move it should receive a nerve impulse passing through 2 main neurones:

1. Upper Motor Neurone (U.M.N.)
2. Lower Motor Neurone (L.M.N.)

I. UPPER MOTOR NEURONE (U.M.N.)

1. The voluntary motor impulse originates mainly in the large pyramidal Δ cells (Betz cells) of the motor area (area 4) & to a lesser extent in the cells of the premotor area (area 6).
2. The axons of these cells descend in the depth of the cerebral hemisphere in the corona radiata, to pass in the **internal capsule** (genu & ant. 1/3 of post. limb) & continue their descent in the midbrain, pons & medulla.
3. In the lower medulla, 80% of the fibres decussate to descend in the white matter of the **opposite** side of the spinal cord, while the remaining fibres descend directly in the white matter of the same side.

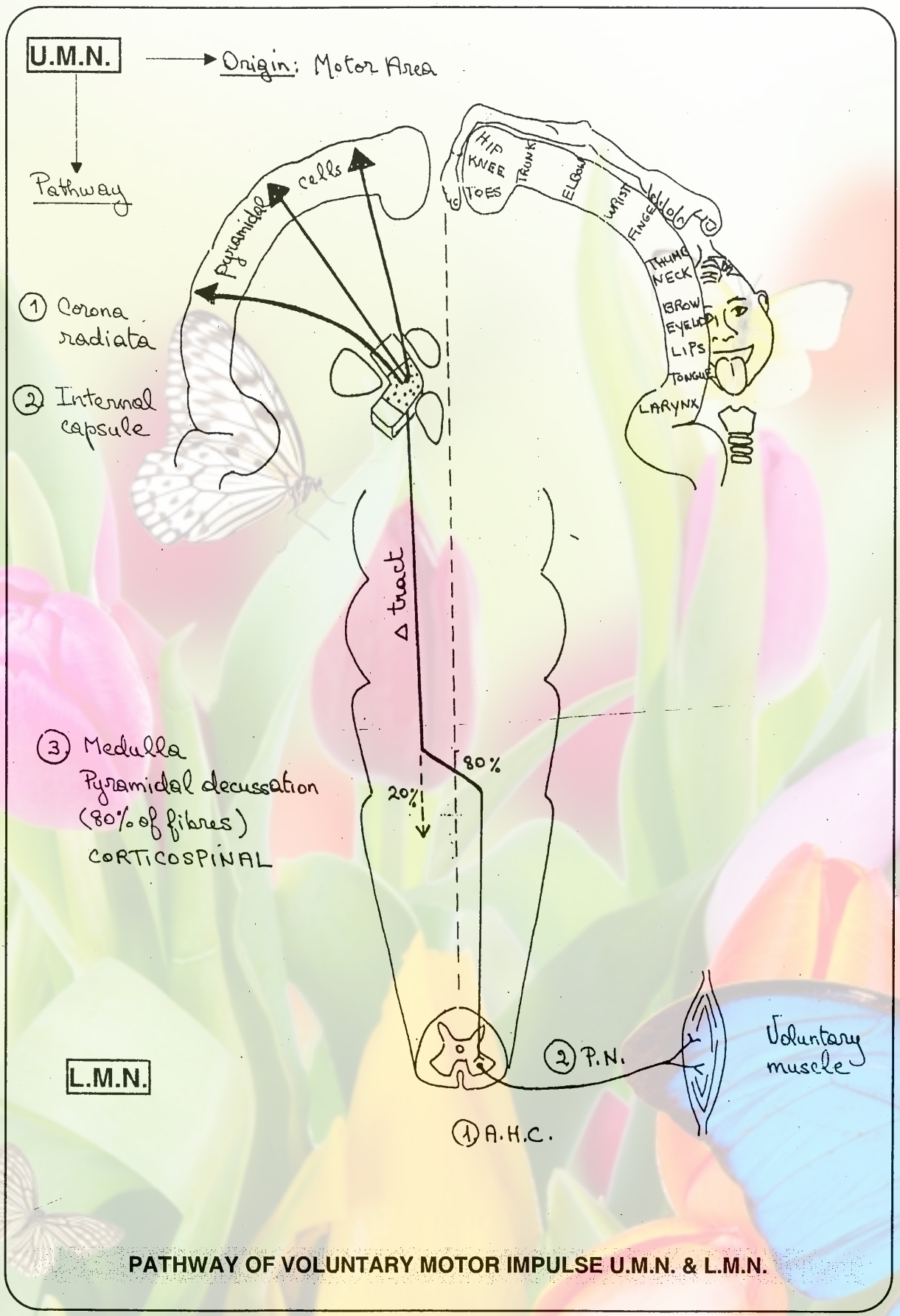
The pathway starting from the cells of the cortex down to the spinal cord is known as the pyramidal (Δ) or Corticospinal Tract.

4. The fibres of the Δ tract terminate at different levels of the A.H.C.s of the spinal cord.
5. In the Brain Stem, some of the descending Δ fibres separate to supply the **motor nuclei of the cranial nerves of BOTH sides except** the lower 1/2 of the facial nucleus & the hypoglossal nucleus which are supplied only from the **opposite** Δ tract. These fibres are known as **corticobulbar** as they do not reach the spinal cord.

II. LOWER MOTOR NEURONE (L.M.N.)

1. **A.H.C.s**: they are a special type of nerve cells situated in the anterior horns of the H-shaped gray matter of the spinal cord. They receive the voluntary motor impulse from the Δ tract. Their axons exit from the spinal cord as the anterior roots.

 N.B.: The **motor nuclei** of the cranial nerves are similar in function to the A.H.C., as they form the cell bodies of the L.M.N. of the cranial nerves. Thus, lesion of a cranial nerve nucleus, like lesion of an A.H.C. is a L.M.N. lesion.
2. **Peripheral Motor Nerve** carrying the motor impulse from A.H.C. to the voluntary muscle.



PATHWAY OF VOLUNTARY MOTOR IMPULSE U.M.N. & L.M.N.

MUSCLE TONE

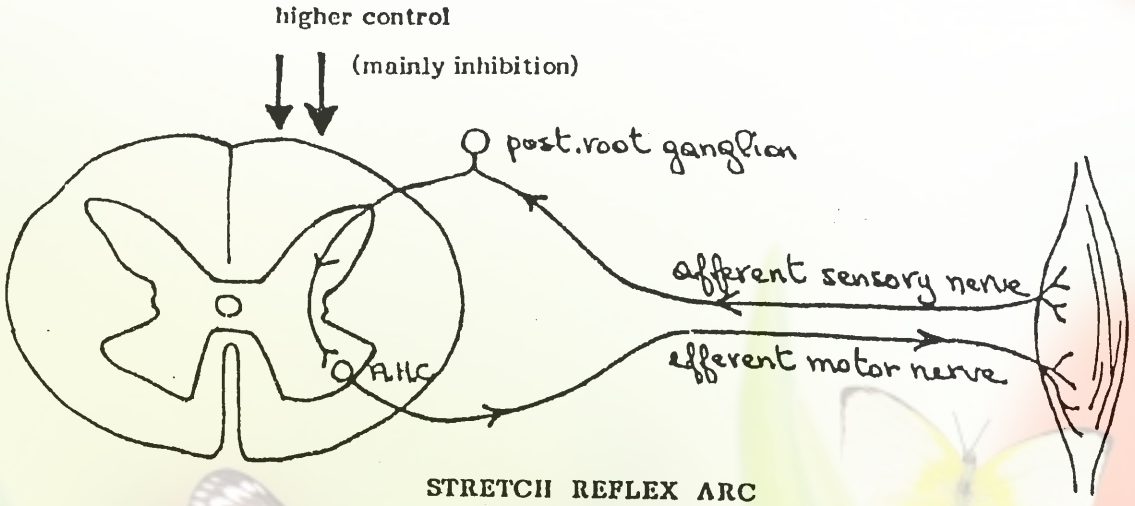
1. This is a **spontaneous local axon stretch reflex**.
2. The length of any skeletal muscle is shorter than the distance between its origin & insertion. This puts the muscle in a state of **constant slight stretch**.
3. This stretch stimulates some muscle spindles which send excitatory impulses through the afferent sensory nerve & the dorsal root to the A.H.C.
4. The excited A.H.C. send motor impulses through the anterior root & the efferent motor nerve to the muscle.
5. This results in continuous reflex subtetatinic contraction of the muscle; this constitutes the Muscle Tone which is important for the nourishment of the muscles & the posture of the body.
6. The Muscle Tone receives higher control, mainly inhibitory, through the pyramidal & extrapyramidal systems. Therefore:
 - **U.M.N.L.** (Δ lesion) results in loss of Δ inhibition of the intact reflex arc leading to increased muscle tone **(spasticity)** below the level of the lesion with no wasting of the muscle.
 - **L.M.N.L.** results in interruption of the reflex arc leading to decreased muscle tone **(flaccidity)** at the level of the lesion, with wasting of the muscles.

TENDON JERK (DEEP REFLEX)

1. This is an **induced local axon stretch reflex**.
2. It is induced by tapping the tendon of the muscle with a hammer. This tap stretches the muscle with synchronous stimulation of all muscle spindles & activation of the local axon reflex (as in Muscle Tone), resulting in a brief contraction of the muscle.
3. The pyramidal system also exerts an inhibitory effect on this stretch reflex. Therefore:
 - **U.M.N.L.** (Δ lesion) results in exaggeration of deep reflexes **(hyperreflexia)** below the level of the lesion.
 - **L.M.N.L.** results in diminution of deep reflexes **(hyporeflexia)** at the level of the lesion.

CLONUS

1. Elicited only when the stretch reflex arc is no longer inhibited (severe Δ lesion).
2. Induced by sudden sustained stretch of muscle tendon, resulting in a series of rhythmic contractions. It stops with relief of tendon stretch.
3. Elicited in ankle, patella & wrist.
4. Clonus may be hysterical where it continues after relief of tendon stretch.



STRETCH REFLEX ARC
(Muscle Tone & Tendon Jerk)

Differnentiation between U.M.N.L. & L.M.N.L.

	U.M.N.L.	L.M.N.L.
1. Paralysis	Paralysis or weakness **below** the level of the lesion.	Paralysis or weakness **at** the level of the lesion.
2. State of muscles	**NO** wasting & if present it is late & due to disuse.	Early & marked **wasting** due to loss of muscle tone.
3. Muscle tone	Hypertonia (**spasticity**) **below** the level of the lesion	Hypotonia (**flaccidity**) **at** the level of the lesion.
4. Fasciculations	Absent.	May be present in **irritative** lesions of **A.H.C.**
5. Deep reflexes	Hyperreflexia **below** the level.	Hyporeflexia **at** the level.
6. Pathological deep reflexes e.g. patellar & adductor reflex	May be present.	Absent.
7. Clonus	May be present.	Absent.
8. Superficial reflexes e.g. abdominals.	Lost if lesion is above the segmental supply of the reflex.	Lost if lesion involves the segmental supply of the reflex.
9. Plantar reflex (Babinski)	**+ ve**, i.e. dorsiflexion of big toe ± fanning of other toes.	Plantar flexion of toes or no response (never say –ve Babinski).

THE SENSORY SYSTEM

Sensations, in general are classified into:

I. **SOMATIC SENSATIONS:** are conducted to the CNS via "somatic nerves;" they include:

1. Superficial sensations:

–Pain –Temperature –Touch.

2. Deep sensations (Proprioceptive):

–Vibration sense –Joint sense –Muscle sense –Nerve sense.

3. Cortical sensations:

–Tactile localisation –2 points discrimination

–Stereognosis –Graphosthesia –Perceptual rivalry

II. **VISCERAL SENSATIONS**: all sensations from internal viscera reaching the CNS via the "autonomic nerves."

III. **SPECIAL SENSATIONS**: including vision, hearing, smell & taste reaching the CNS via the "cranial nerves."

SOMATIC SENSATIONS

1. All somatic sensations, whether superficial or deep pass through **3 order neurones** from receptors in the skin & deep structures to reach the cortical sensory area of the opposite side.
2. The cell of the 1st order neurone is always in the **posterior root ganglion.**
3. The cell of the 3rd order neurone is always in the **thalamus** of the **opposite** side.
4. The 2nd order neurone varies according to the type of sensation.

A) PATHWAY OF SUPERFICIAL SENSATIONS

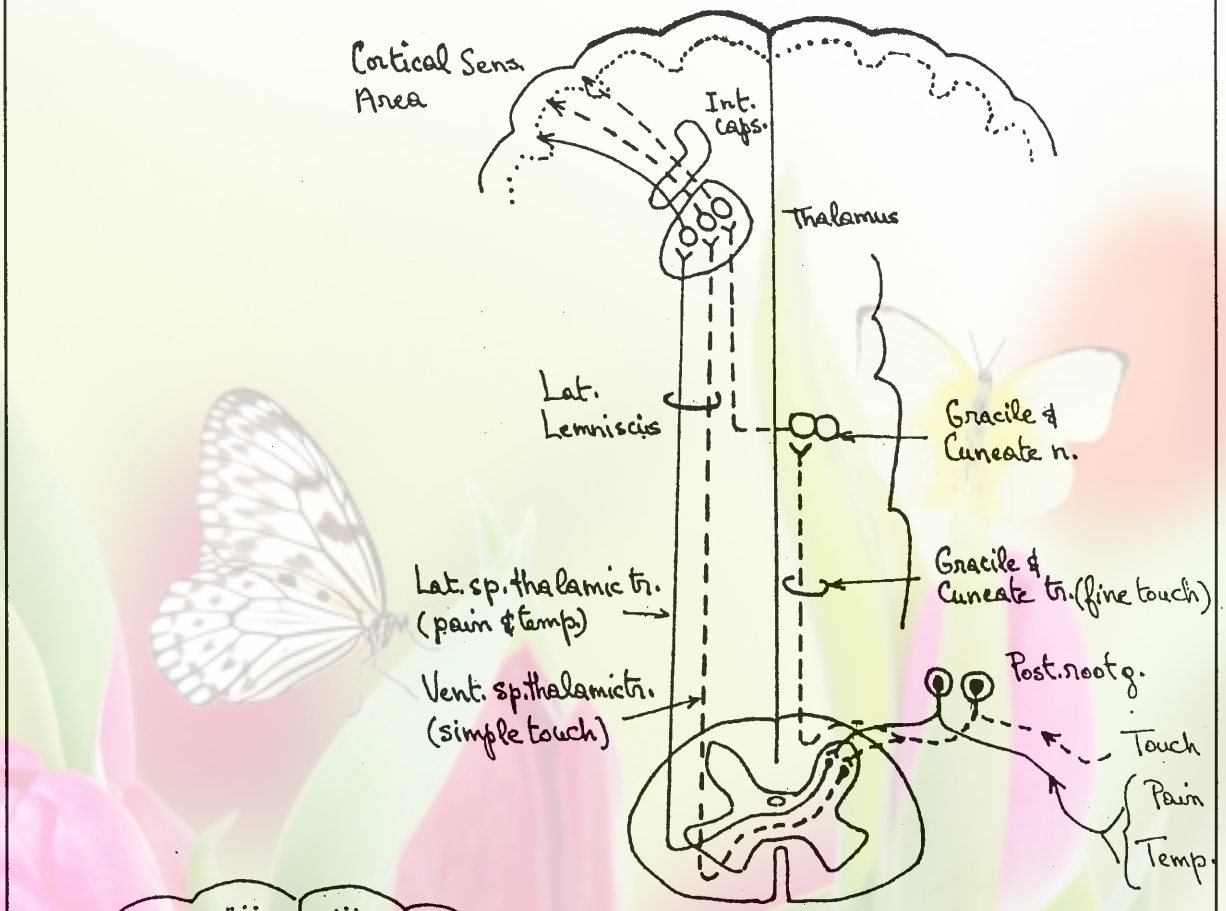
I. Pathway of Pain & Temperature

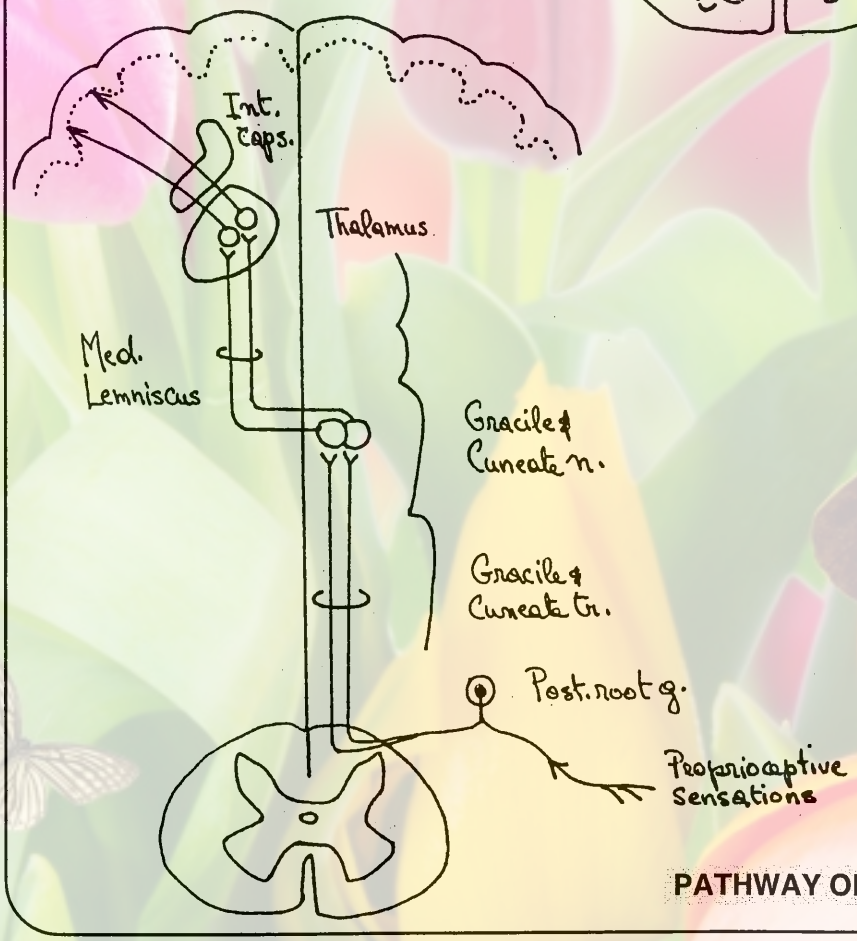
The 1st order neurone is the cell of the **posterior root ganglion** & its axon. This axon is divided into a lateral & a medial branch. The lat. branch forms the afferent sensory nerve. The medial branch enters the spinal cord to ascend a few segments forming **Lissauer's tract**, and relays in the cells of Substantia Gelatinosa of Rolandi **(S.G.R.)** capping the post. horn of the gray matter.

The 2nd order neurone is the cell of **S.G.R.** & its axon. This axon crosses to the **OPPOSITE** side & ascends in the **Lateral Spinothalamic Tract** of the spinal cord then in the lateral lemniscus of the brain stem, to relay in the thalamus.

The 3rd order neurone starts in the cell of the **thalamus**, its axon ascends to pass through the posterior limb of the internal capsule conducting the impulse to the cortical sensory area in the parietal lobe.

PATHWAY OF SUPERFICIAL, SENSATIONS





PATHWAY OF DEEP SENSATIONS

II. Pathway of Touch

1. **Simple** touch has the same pathway as pain & temp. but in the 2nd order neurone, it ascends in the **Ventral Spinothalamic** tract of the **opposite** side of the spinal cord → Lateral Lemniscus → Thalamus → Cortical Sensory Area (1, 2, 3).
2. **Fine** touch has the same pathway as deep sensations.

B) PATHWAY OF DEEP (PROPRIOCEPTIVE) SENSATIONS

The 1st order neurone is the cell of the **posterior root ganglion** & its axon which is divided into a lateral & medial branch. The lateral branch forms the afferent sensory nerve. The med. branch enters the spinal cord & ascends in the **Gracile & Cuneate tracts** within the post. column of the **SAME** side (along with fibres carrying fine touch) to relay in the Gracile & Cuneate nuclei in the medulla:

* Gracile Tract: – carries fibres from lower $^1/_2$ of body and lies medially.
* Cuneate Tract: – carries fibres from upper $^1/_2$ of body and lies laterally.

The 2nd order neurone from the cell of the **Gracile & Cuneate nuclei**, the axon crosses to the opposite side & ascends in the **Medial Lemniscus** through the brain stem to relay in the thalamus.

The 3rd order neurone the same as in superficial sensations.

C) CORTICAL SENSATIONS

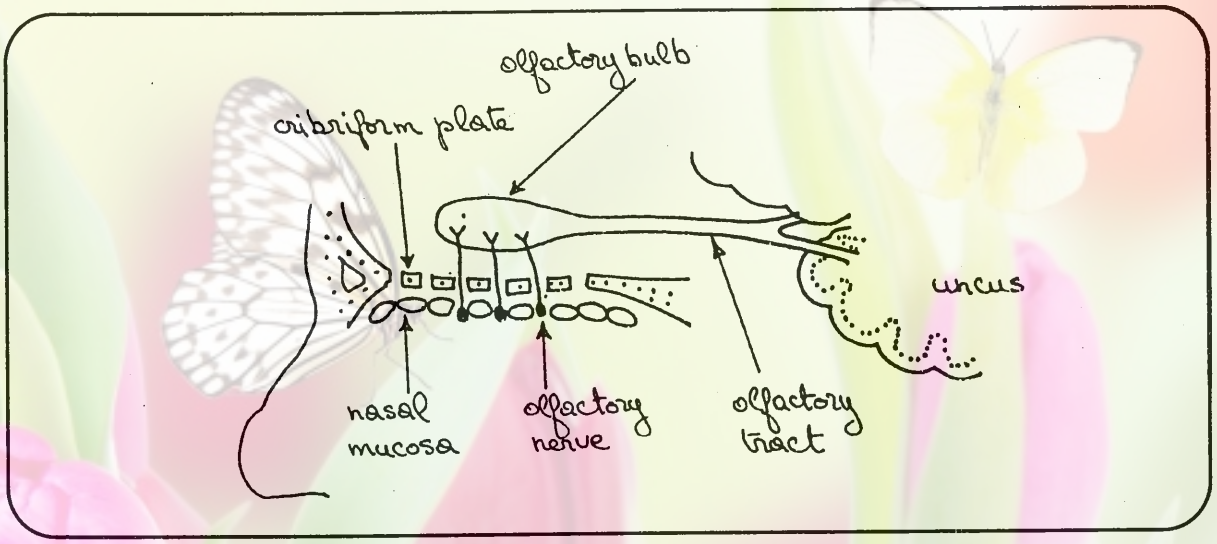
1. These are a mixture of refined superficial & deep sensations arriving to the thalamus via the 1st & 2nd order neurones & conducted from the thalamus, via the 3rd order neurone to the cortical sensory area (1, 2, 3) in the parietal lobe.
2. They allow the perception of the exact localisation of a stimulus **(tactile localisation, 2-point discrimination)** & the perception of the variations of forms & textures of objects **(stereognosis . . .)**.

THE CRANIAL NERVES

I. THE OLFACTORY NERVE

PATHWAY OF SMELL:

From the receptors in the olfactory mucosa the fibres of the olfactory nerve pierce the cribriform plate of the ethmoid bone and run in the olfactory groove to relay in the olfactory bulb. A new set of fibres travels in the olfactory tract to terminate in the olfactory sensory area in the uncus of the temporal lobe of both sides.



LESIONS:

I. **ANOSMIA**: Loss of sense of smell, it may be unilateral or bilateral.

A. **Unilateral anosmia**: Causes:

1. Traumatic: Fracture cribriform plate (as in fracture base of skull).
2. Inflammatory: Basal meningitis.
3. Neoplastic: – Olfactory groove meningioma, – Tumours of inf. Surf. Or frontal lobe } Foster-Kennedy syndrome

* **Foster-Kennedy syndrome**: These tumours cause:

- ipsilateral 1ry optic atrophy due to direct pressure on the optic nerve,
- contralateral papilloedema due to ↑ intracranial tension (↑ I.C.T.),
- ipsilateral anosmia may occur due to pressure on the olfactory pathway.

N.B.: Unilateral lesion of the uncus does not cause anosmia as the sense of smell is bilaterally represented.

B. **Bilateral anosmia**: Causes:

1. E.N.T. causes as common cold or atrophic rhinitis.
2. Congenital. 3. Hysterical.

II. **PAROSMIA**: Perverted sense of smell. Strong scents, e.g. perfumes, smell abnormal, usually unpleasant. Commonest cause is Post-traumatic.

III. **OLFACTORY HALLUCINATIONS**: Perception of smell usually unpleasant, in the absence of stimulus. It is due to an irritative lesion in or near the uncus. It is usually part of temporal lobe epilepsy.

2. THE OPTIC NERVE

PATHWAY OF VISION:

The receptors for vision are the cones (for day vision) and rods (for night vision) of the retina. From these receptors the nerve fibres converge and pass through the optic disc to form the optic nerve which contains nasal and temporal fibres. The nasal fibres decussate at the optic chiasma to pass in the optic tract of the opposite side, while the temporal fibres pass in the optic tract of the same side. The fibres of the optic tract relay in the lateral geniculate body. A new set of fibres arises and passes in the posterior limb of the internal capsule, then fans out as the optic radiation where the upper fibres run in the parietal lobe while the lower fibres run in the temporal lobe. The fibres of the optic radiation finally terminate in area 17 of the occipital lobe (visual sensory area). The recognition and identification of objects is achieved in the neighbouring areas 18 and 19 (visual psychic areas)

LESIONS OF THE OPTIC NERVE:

A. Lesion in the Optic Nerve:

1. Ipsilateral loss of vision with 1ry optic atrophy.
2. Loss of direct and consensual light reflex, on exposure of the affected eye to light.

B. Lesion in the Optic Chiasma:

1. From above e.g. suprasellar tumour produces lower quadrantic bitemporal hemianopia.
2. From below e.g. intrasellar tumour produces upper quadrantic bitemporal hemianopia.
3. Complete destruction of the chiasma produces bitemporal hemianopia.

C. Lesion of the Optic Tract:

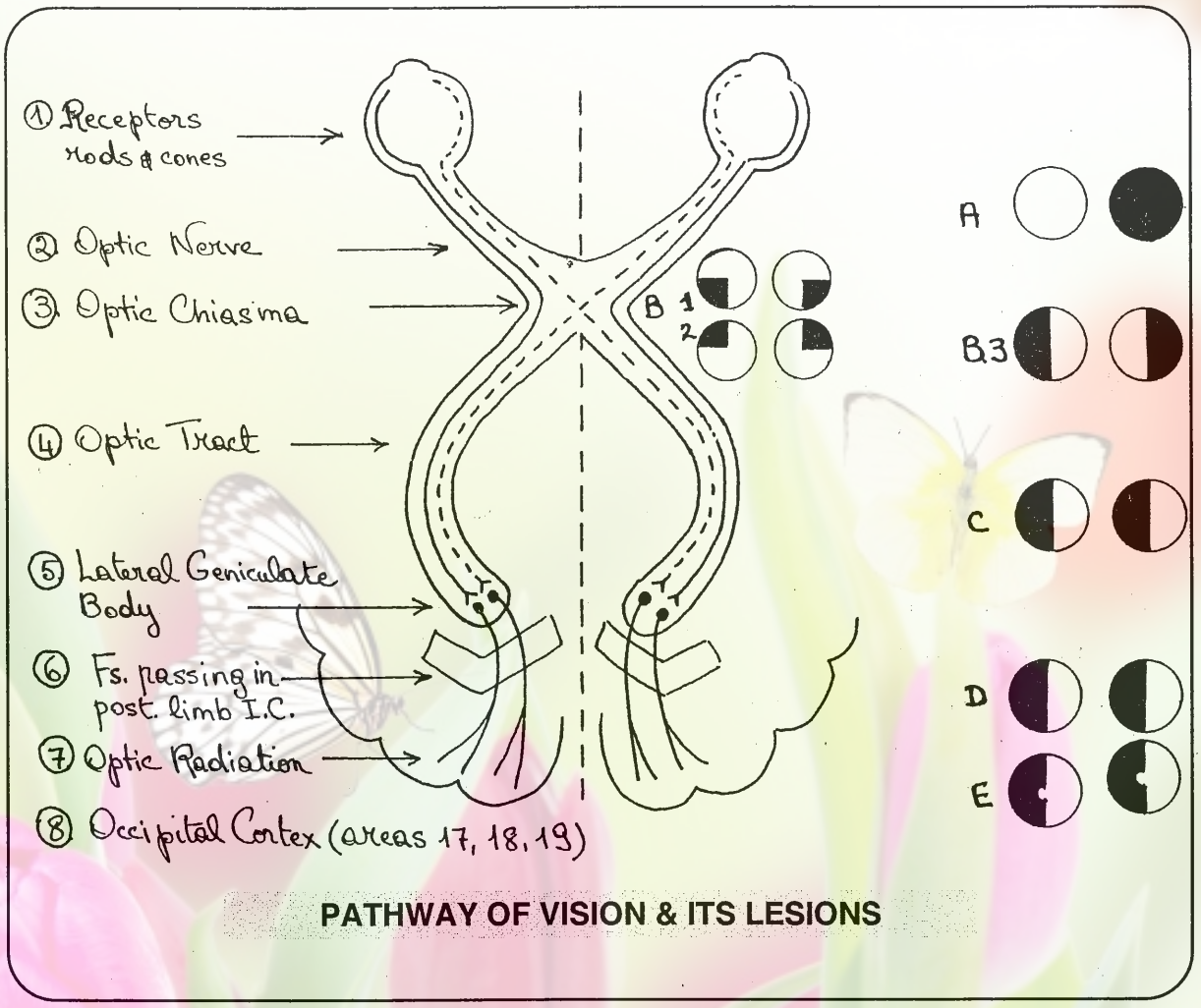
1. Contralateral homonymous hemianopia.
2. Hemianopic pupillary reaction: exposure of the nasal (seeing) side of the retina of the affected eye results in a pupillary reaction; while exposure of the temporal (non-seeing) side results in no pupillary reaction.

D. Lesion of the Optic Radiation:

1. Contralateral homonymous hemianopia.
2. The light reflex is preserved.

E. Lesion of the Occipital Lobe:

1. Contralateral homonymous hemianopia with or without macular sparing. The macular presentation on the occipital cortex has double blood supply (from the posterior and middle cerebral arteries); thus in vascular lesions there is macular sparing.
2. Preservation of the light reflex.



PATHWAY OF VISION & ITS LESIONS

3. THE OCULOMOTOR NERVE

The nucleus of this nerve lies in the midbrain anterior to the aqueduct of Sylvius & close to the midline.

It has a complex structure & is formed of several nuclei of different functions:

- Motor nuclei to the extraocular muscles.
- Parasympathetic nuclei: - Edinger-Westphal nucleus to the constrictor pupillae muscle.
 - Perlia's nucleus to the ciliary muscle.

The oculomotor nerve passes forwards through the cavernous sinus to enter the superior orbital fissure & supply the following muscles of the eye.

1. **Extraocular muscles**: medial, superior & inferior recti, inferior oblique & levator palpebrae superioris (responsible for 70% of elevation of the upper eyelid).
2. **Intraocular muscles**: – Constrictor pupillae muscle of the iris.
 – Ciliary muscle of the lens.

LESION:

A. External Ophthalmoplegia:

1. Ptosis due to paralysis of the levator palpebrae muscle.
2. Divergent paralytic squint, the eye looks out & down due to unopposed action of the lateral rectus (Cr 6) & the superior oblique (Cr 4) muscles.
3. Diplopia on passive elevation of the eyelid.

B. Internal Ophthalmoplegia:

1. Ipsilateral dilated fixed pupil (mydriasis).
2. Affection of the light reflex: on exposure of the eye on the affected side to bright light the direct light reflex is lost while the consensual reflex is preserved.

N.B. :In the oculomotor nerve the autonomic fibres lie superficial to the motor fibres.
∴ **Compression** of the nerve e.g. by a (tumour) results in affection of the superficial fibres with **early mydriasis & lost light reflex.**
While **infarction** of the nerve e.g. by (ischaemia, diabetes) results in sparing of the superficial fibres & the **pupillary reflexes remain intact.**

4. THE TROCHLEAR NERVE

The nucleus of this nerve lies in the lower part of the midbrain. The nerve supplies the Superior Oblique muscle which turns the eye inwards & downwards.

LESION:

1. **Diplopia** only when the patient looks downwards e.g. when descending the stairs or reading.
2. **Limitation of movement** of the affected eye on looking inwards & downwards.

6. THE ABDUCENT NERVE

The nucleus of this nerve lies in the lower part of the pons. The nerve runs a long intracranial course to supply the lateral rectus muscle which moves the eye outwards (laterally).

LESION: It is the most commonly affected ocular nerve because of its long course.

1. **Diplopia** only when the patient looks outwards, towards the paralysed side.
2. **Limitation of movement** of the affected eye on looking outwards.

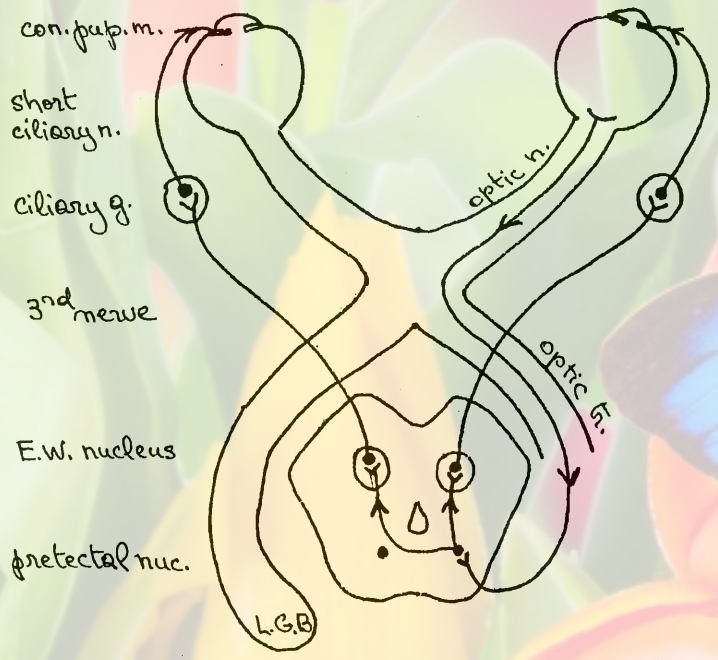
LIGHT REFLEX:

Exposure of one eye to bright light (with shading of other eye) results in constriction of the pupil of the same side (direct reaction) & of the opposite side (consensual reaction).

PATHWAY:

1. Exposure of the eye to bright light sends impulses along the optic nerve, chiasma and tract.
2. The fibres concerned with the light reflex do not reach the lateral geniculate body but pass from the optic tract to the pretectal nucleus in the midbrain where they relay.
3. New fibres pass to the Edinger-Westphal nucleus (part of Cr III nucleus) of both sides; the decussating fibres pass around the aqueduct of Sylvius & account for the consensual reaction to light.
4. From the E.W. nucleus of both sides preganglionic fibres pass through the oculomotor nerves to relay in the ciliary ganglia.
5. Postganglionic fibres pass by the short ciliary nerves to the constrictor pupillae muscles.

Optic nerve → Optic tract → Pretectal nucleus → E.W. nucleus (both sides) → 3rd nerves → Ciliary ganglia → Short ciliary nerves → Constrictor pupillae muscles



PATHWAY OF LIGHT REFLEX

ACCOMMODATION (NEAR) REFLEX:

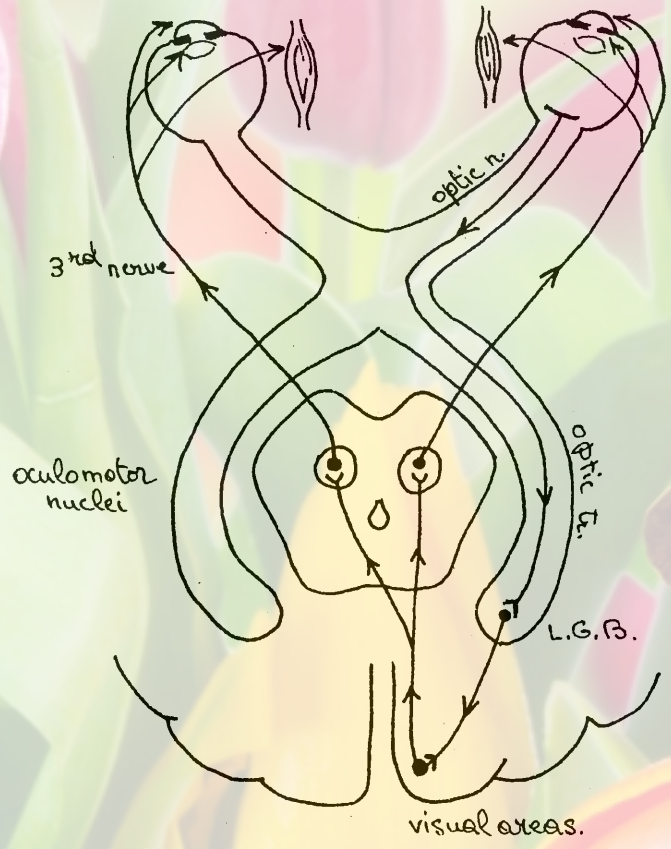
When the eyes focus on a near object, there is the following triad:

1. **Convergence** of the eyes due to contraction of both medial recti muscles.
2. **Miosis** (constriction of the pupils) due to contraction of the constrictor pupillae muscles.
3. **Accommodation** (increased refractive power of the lens) due to contraction of ciliary muscles.

PATHWAY

1. Afferent impulses from the retina pass along the normal visual pathway to reach the visual areas in the occipital lobe.
2. From the visual areas fibres descend to the oculomotor nuclei of both sides in the midbrain.
3. Efferent fibres pass along the oculomotor nerves to the eyes to supply:
 * Medial recti muscles. * Constrictor pupillae muscles. * Ciliary muscles.

In this reflex, the fibres reach the lat. geniculate body & the occipital cortex but do not pass through the pretectal nucleus in the midbrain.



PATHWAY OF ACCOMMODATION REFLEX

THE PUPIL

The size of the pupil is affected by 2 groups of smooth muscles acting on the iris.

1. Constrictor pupillae muscle which is dominant & receives parasympathetic supply from the oculomotor nerve & reaching it via the short ciliary nerves.
2. Dilator pupillae muscle which receives sympathetic supply via the long ciliary nerves.

PATHWAY OF SYMPATHETIC SUPPLY:

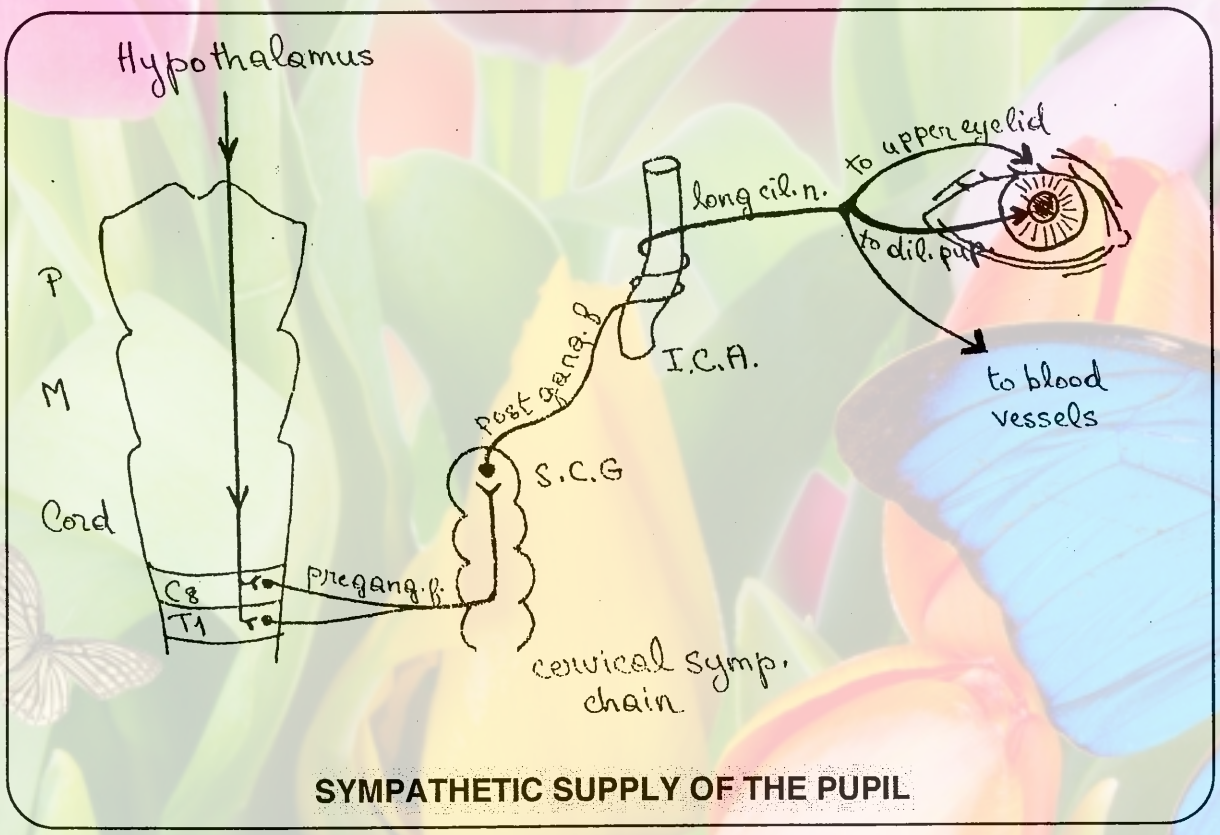
It starts from the hypothalamus and descends through the brain stem & spinal cord to the lateral horn cells of C8 & Th1 segments. Preganglionic fibres pass in the anterior roots of C_8 & Th_1 nerves and ascend in the cervical sympathetic chain to relay in the superior cervical ganglion. Post-ganglionic fibres ascend around the internal carotid artery to enter the cranial cavity. They then leave the artery and pass mainly through the long ciliary nerves to supply:

1. Dilator pupillae muscle.
2. Smooth muscles of the upper eyelid, responsible for 30% of its elevation.
3. Vasoconstrictive fibres to the eye & face.

N.B.: Sympathetic fibres concerned with sweating ascend from the superior cervical ganglion, with the external carotid artery to supply the sweat glands of the face.

SPINO-CILIARY REFLEX

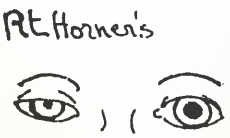
Pinching the skin on one side of the neck results in dilatation of the ipsilateral pupil. The efferent limb of this reflex is the sympathetic supply to the pupil.



SYMPATHETIC SUPPLY OF THE PUPIL

PUPILLARY DISORDERS:

A) MYOSIS: Causes:



1. Horner's syndrome (Sympathetic lesion):

1. Ptosis (partial).
2. Miosis.
3. Enophthalmos.
4. Conjunctival congestion
5. Lost spino-ciliary reflex.
6. Anhydrosis.

2. Argyll-Robertson pupil:

1. Myotic irregular eccentric pupil.
2. Reactive to accommodation but not to light.
3. Sluggish response to the spino-ciliary reflex and to mydriatics.
4. It is due to a lesion in the periaqueductal region of the midbrain where the fibres of the light reflex are close to the sympathetic pupillodilator fibres.
5. Causes:
 - Neurosyphilis.
 - Encephalitis.
 - D.S.
 - Diabetes mellitus.
 - Syringobulbia.
 - Chronic alcoholism.



3. Pontine haemorrhage.
4. Opiate overdosage.
} pin-point pupil

B) MYDRIASIS: Causes



1. **Oculomotor Nerve Lesions:** Dilated fixed pupil.

2. **Marcus-Gunn Pupil:** In early optic nerve lesion where there is affection of the afferent limb of the light reflex: if a light source is " swung" from eye to eye, lasting 4 seconds on each, the affected pupil may paradoxically dilate (a Marcus-Gunn pupil).



3. **Adie's Pupil:**
 1. The pupil is dilated, with no reaction to light.
 2. Slow reaction to accommodation & on relaxation of accommodation, the pupil dilates very slowly (tonic pupil).
 3. It may be associated with lost ankle reflex.
 4. It is usually unilateral, of acute onset, occurring in females & is benign.



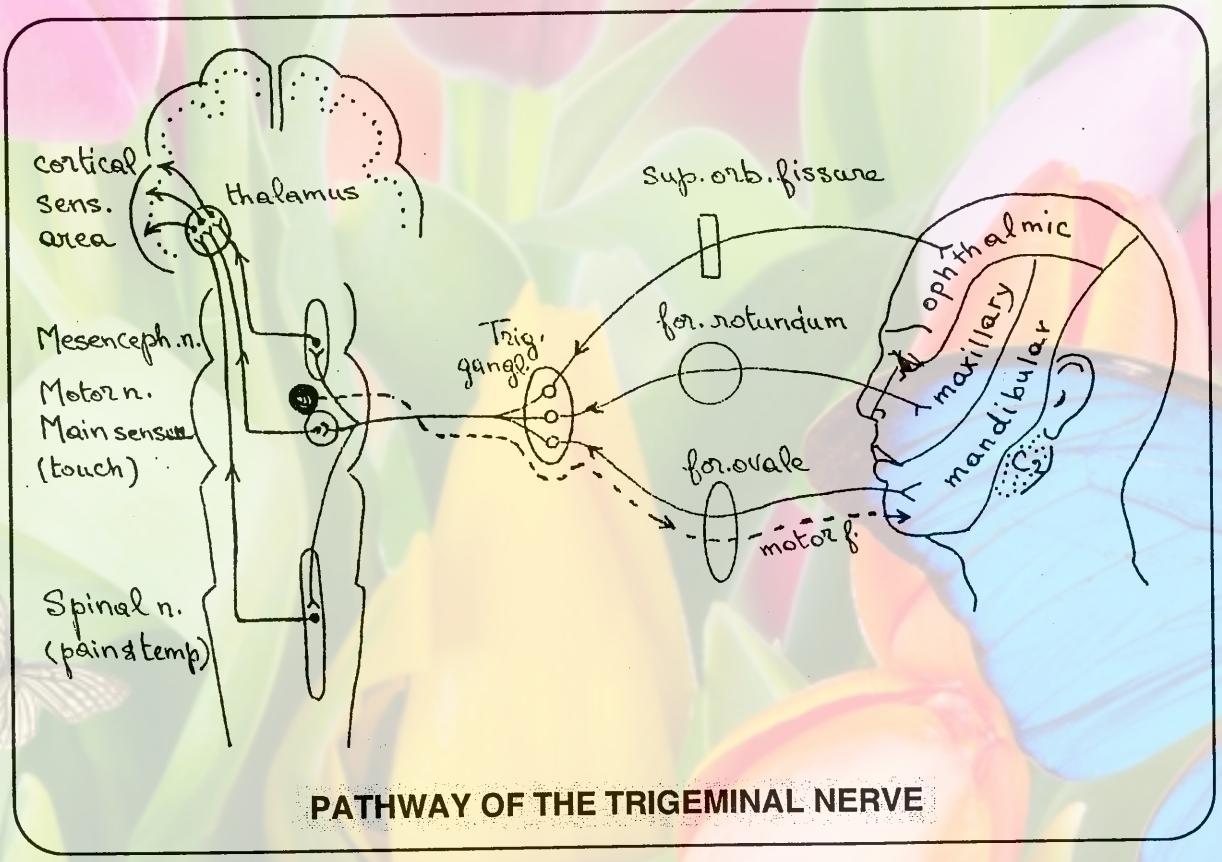
5. THE TRIGEMINAL NERVE

It is formed of a sensory & a motor division.

1) <u>The Sensory Division:</u> It conducts sensations from the face (except the angle of the mandible supplied by C2), the anterior $^2/_3$ of the tongue & the buccal cavity.

It is formed of 3 branches, the **ophthalmic**, **maxillary** and **mandibular** branches which enter the cranial cavity respectively through the superior orbital fissure, the foramen rotundum and the foramen ovale to terminate in the Trigeminal (Gasserian) Ganglion. This ganglion contains bipolar cells; their central branches pass into the pons, where they take one of 3 pathways:

1. Fibres carrying **pain and temperature** sensations run downwards to relay in the **spinal nucleus** in the lower part of the medulla on the same side. In this nucleus:
 - The face is represented upside down i.e. ophthalmic fibres end in the lower part and mandibular fibres in the upper part.
 - The peripheral part of the face is represented in the inner part of the nucleus and the central part of the face is represented in its outer part.
2. Fibres carrying **proprioceptive** sensations run upwards and relay in the **mesencephalic nucleus** in the midbrain, on the same side.
3. Fibres carrying **touch** pass directly to the **main sensory nucleus** in the pons on the same side.

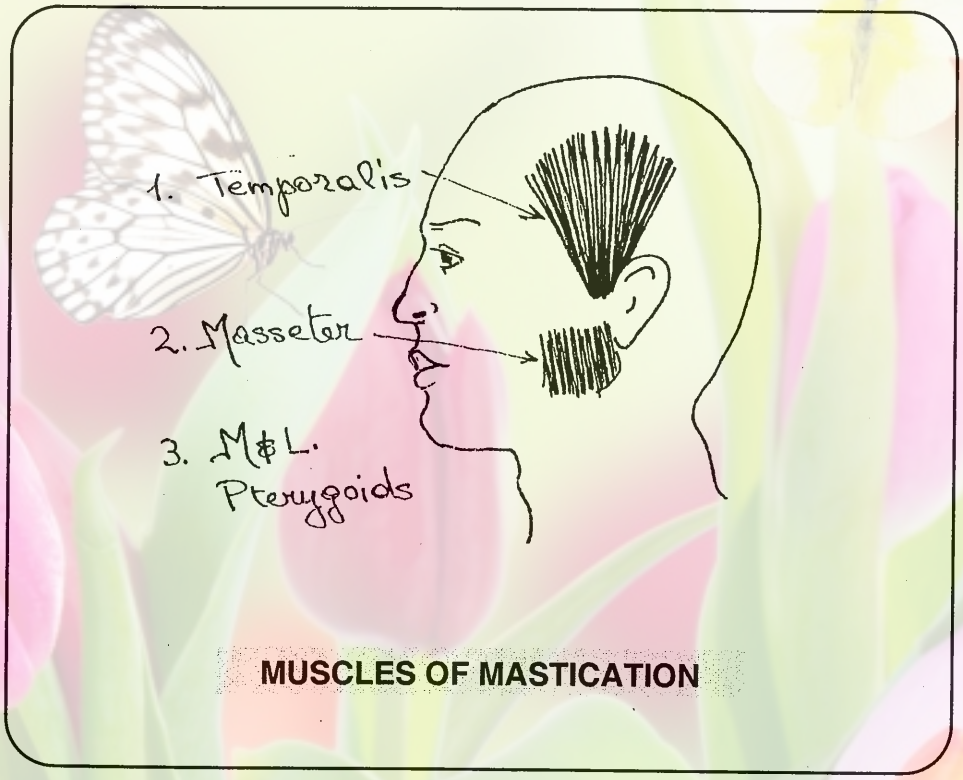


PATHWAY OF THE TRIGEMINAL NERVE

From these 3 nuclei new fibres cross to the opposite side, then ascend with the medial and lateral lemnisci, to reach the thalamus.
From the thalamus, fibres carrying cortical sensations of the face ascend to end in the cortical sensory area of the face.

2) The Motor Division: It starts in the motor nucleus in the pons. The fibres emerge from the pons & pass laterally below the trigeminal ganglion to join the mandibular branch of the nerve which leaves the cranial cavity through the foramen ovale. It supplies:

1. **The muscles of mastication**: Temporalis, Masseter, Lateral & Medial Pterygoids.
2. Anterior belly of digastric: 3. Mylohyoid. 4. Tensor palati.



MUSCLES OF MASTICATION

LESIONS OF THE TRIGEMINAL NERVE:

A) Sensory Affection:

1. Peripheral Lesion: Loss of sensations on the same side of the face (sparing the angle of the mandible), including all of the face or the area supplied by the affected branch.
 In mandibular lesions there is also loss of general sensations over anterior $^2/_3$ of tongue.
2. Central Lesion: (mainly affecting the Spinal Sensory, nucleus in the medulla) resulting in ipsilateral **dissociated sensory loss** of the face i.e. lost pain & temperature with sparing of touch & deep sensations. The clinical picture varies:

Lesion starting from above (lower pontine tumour)	→ affection of lower part of face.
Lesion starting from below (upper cervical lesions)	→ affection of upper part of face.
Lesion starting from periphery (tabes dorsalis)	→ affection of central part of face.
Lesion starting from midline (syringobulbia)	→ affection of peripheral part of face with sparing of the central (muzzle) area.

B) Motor Affection:

1. Weakness of the muscles of mastication on the same side of the lesion.
2. Deviation of the jaw to the affected side due to the unopposed action of the pterygoid muscles of the healthy side.

C) Reflex Affection:

1. Ipsilateral loss of the corneal & conjunctival reflexes (afferent 5, efferent 7).
2. Ipsilateral loss of the palatal reflex (afferent 5, efferent 10).
3. Exaggerated jaw reflex (afferent 5, efferent 5) in cases of bilateral U.M.N.L. above the pons.

D.D. OF FACIAL PAIN

1. Trigeminal neuralgia (Tic douloureux):

- Severe attacks of unilateral pain along one or more of the sensory branches of the trigeminal nerve, usually the mandibular or maxillary.
- It usually affects middle age, more commonly females.
- The exact cause is unknown but there are certain predisposing factors:
 - Root compression by a tumour or blood vessels in the cerebello-pontine angle.
 - Trigeminal neuralgia is common in D.S., diabetes and in alcoholism.
- The attacks are precipitated by movements of the jaw as laughing, brushing of the teeth, mastication ... & last several days or weeks.
- In between attacks the patient is completely free.
- Treatment:

 a) Medical:
 - Carbamazepine (Tegretol) 600-1200mg/day or Clonazepam (Rivotril) 2-6 mg/d.
 - Analgesics e.g.: Acupan or Idarac may be used.

 b) Surgical:
 - Injection of nerve or ganglion with alcohol or phenol provides relief for up to one year.
 - Section of the affected sensory root.
 - Microvascular decompression: Sometimes blood vessels in the C.P. angle adhere to and press on the trigeminal nerve roots; the separation of these structures and the insertion of a non-absorbable sponge between them provides pain relief without the side effects of nerve destruction.

2. Post-herpetic neuralgia: the pain usually affects the ophthalmic division; there is evidence of scarring.
3. Sinusitis: the pain is worse in the morning with tenderness over involved sinus.
4. Ocular (glaucoma): pain behind the eye with associated visual symptoms.
5. Dental: pain around the mouth.
6. Atypical facial pain: pain is diffuse & persistent and is associated with depression.
7. Cluster headache: pain above & behind the eye associated with lacrimation/rhinorrhea.
8. Costen's syndrome: temporomandibular joint dysfunction the pain is in front of the ear and is aggravated by chewing.

7. THE FACIAL NERVE

The facial nerve is a mixed nerve, as it contains motor, sensory and autonomic fibres.

The **motor** part supplies the muscles of expression of the face as well as 4 other muscles:

- Platysma.
- Posterior belly of the digastric muscle.
- Stapedius.
- Stylohyoid.

The **sensory** part receives taste sensations from the anterior $^2/_3$ of the tongue.

The **autonomic** part supplies the lacrimal gland as well as the submaxillary and sublingual salivary glands.

Anatomy of the motor part:

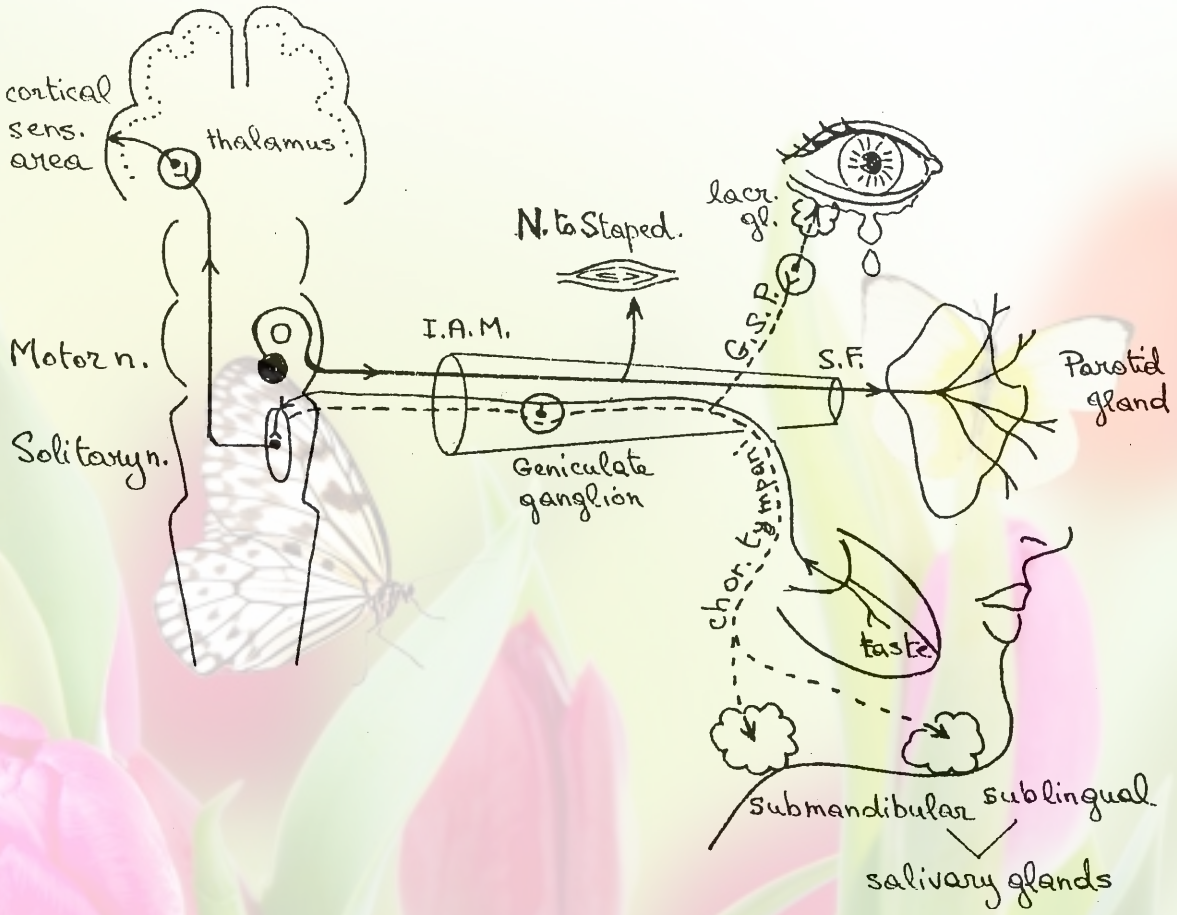
The motor nucleus of the facial nerve is located in the **pons**, medial to the trigeminal nucleus and anterior to the 6th nerve nucleus. Its upper part is bilaterally supplied from the pyramidal tracts of both sides, while the lower part is unilaterally supplied from the pyramidal tract of the opposite side only. From the nucleus, the motor fibres form a loop around the 6th nerve nucleus, then pass laterally to emerge at the lower part of the pons. The nerve runs laterally between the 6th and 8th cranial nerves, in the subarachnoid space of the **cerebello-pontine angle** to enter, through the **internal auditory meatus**, into the facial canal. In the **facial canal**, the motor part of the facial nerve becomes adherent to its sensory and autonomic parts. It then leaves the canal through the **stylomastoid foramen**, passes through the **parotid gland** to divide into its terminal branches.

Anatomy of the sensory and autonomic parts:

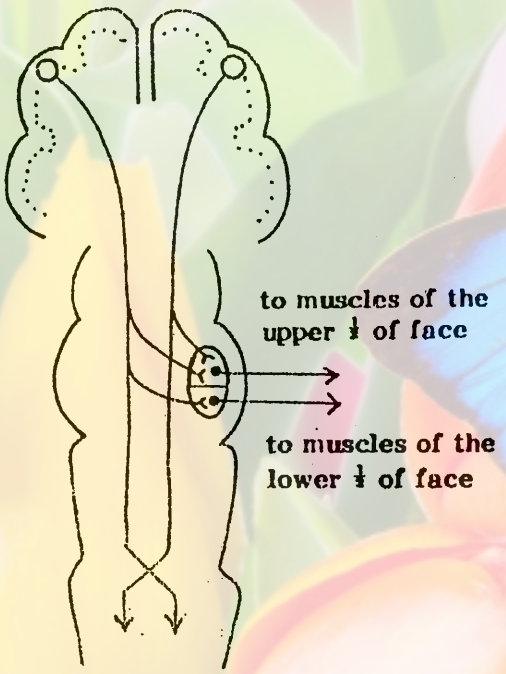
In the facial canal lies the **geniculate ganglion**, which contains unipolar cells. The process of these cells divides in a T-shaped manner into a peripheral branch and a central branch.

1. **The peripheral branch** runs laterally and divides into the greater superficial petrosal nerve and the chorda tympani.
 - The **greater superficial petrosal** nerve passes forwards to relay in the sphenopalatine ganglion where a new set of fibres gives autonomic supply to the lacrimal gland.
 - The **chorda tympani** leaves the facial nerve before the stylomastoid foramen, to supply the submaxillary and sublingual salivary glands and to carry taste sensations from the anterior $^2/_3$ of the tongue.
2. **The central branch** of the unipolar cells passes centrally, joins the motor part of the nerve, then enters the cranial cavity through the internal auditory meatus as the **nervus intermedius**. The latter enters the brain stem at the pontomedullary junction to terminate in the **solitary nucleus** in the medulla. A new set of fibres passes from the nucleus to the opposite side and runs upwards to terminate in the lower part of the **cortical sensory area**, where taste sensation from the ant. $^2/_3$ of the tongue is perceived.

MOTOR, SENSOR & AUTONNOMIC PARTS OF THE FACIAL NERVE



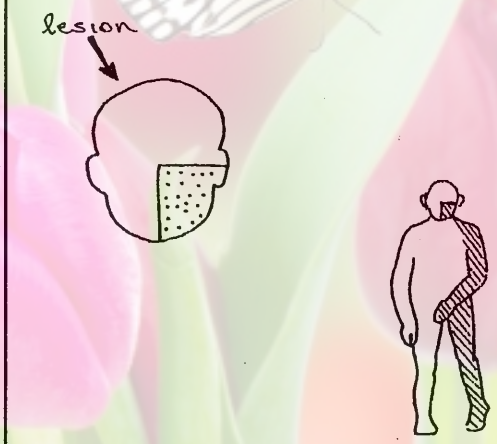
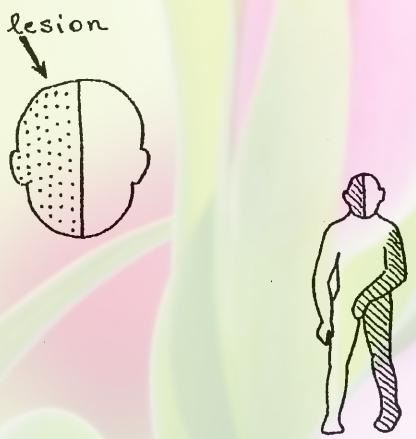
PYRAMIDAL SUPPLY TO THE FACIAL NUCLEUS



LESIONS OF FACIAL NERVE

The lesion may be:

1. U.M.N.L. affecting the pyramidal tract above the facial nucleus.
2. L.M.N.L. affecting the facial motor nucleus or the nerve itself.

U.M.N.L	L.M.N.L.
1. Affecting Δ tract above facial nucleus.	1. Affecting the facial motor nucleus or the nerve itself.
2. Paralysis of the muscles of lower half of the face on the opposite side of the lesion (supplied from the opposite Δ tract only).	2. Paralysis of the muscles of the upper & lower halves of the face on the same side of the lesion.
3. Paralysis involves the voluntary movement; it spares the emotional & associative movements (which are supplied by extra Δ fibres).	3. Paralysis affects voluntary, emotional & associative movements.
4. Paralysis is associated with hypertonia & hyperreflexia.	4. Paralysis is associated with hypotonia & hyporeflexia.
5. There is associated hemiplegia on the same side of the facial paralysis.	5. If there is hemiplegia, it is on the opposite side of the facial paralysis (crossed hemiplegia).
	

N.B.: • In U.M.N. facial paralysis there is, on the affected side:

1. Obliteration of the naso-labial fold.
2. Dropping of the angle of the mouth with dribbling of saliva.
3. Accumulation of food behind the cheek.
4. Inability to blow the cheek.
5. Inability to show the teeth properly.

• In L.M.N. facial paralysis there are, in addition:

6. Inability to raise the eyebrows with absence of wrinkles of the forehead.
7. Inability to close the eye, when the pt. attempts to close his eye the eyeball rolls upwards (Bell's phenomenon).

All above 7 criteria are present in Bell's Palsy (See p 30).



LOCALISATION OF THE SITE OF LESION IN L.M.N. FACIAL PARALYSIS

SITE OF LESION	CAUSES
Nuclear Lesion (in pons): 1. Paralysis of facial muscles. 2. No impairment of taste sensation. 3. No impairment of salivation nor lacrimation. 4. May be other L.M.N. cranial palsies on same side or hemiplegia on opposite side.	1. Vascular: - Vertebro-basilar insuf. - Millard-Gubler syndrome. 2. Infective: - Encephalitis. - Poliomyelitis. 3. Neoplastic: - Astrocytoma - Glioma 4. Demyelinating: - D.S. - D.E.M.
Cerebello-Pontine Angle Lesion: 1. Paralysis of facial muscles. 2. Diminished taste on ant. $^2/_3$ of tongue. 3. Diminished salivary & lacrimal secretions. 4. Associated Cr 5, 6 & 8 palsies on same side.	1. Infective: Basal meningitis. 2. Neopl.: - Acoustic neuroma. - Meningioma
Facial Canal Lesion: 1. Paralysis of facial muscles. 2. Diminished taste on ant. $^2/_3$ of tongue & diminished salivation if chorda tympani is involved. 3. Diminished lacrimation if the greater superficial petrosal nerve is involved. 4. Hyperacusis if the nerve to Stapedius is involved.	1. Traumatic: - Fracture base. 2. Infective: – Otitis media. – Herpes zoster. 3. Neoplastic: – Facial neuroma. 4. Bell's palsy.
Extracranial Facial Lesion: After its exit from the stylomastoid foramen: Paralysis of facial muscles ***only***.	1. Neuropathy: – Diabetic – Leprotic – Ac. Infective. 2. Myopathy: Facioscapulo-humeral. 3. Myotonia Atrophica. 4. Myasthenia. 5. Invasion by a tumour e.g. from parotid. 6. Injury to the nerve during surgery.

CAUSES OF FACIAL NERVE PARALYSIS:

1. Causes of U.M.N. facial paralysis: same causes of hemiplegia, above the level of the pons (p. 39).
2. Causes of L.M.N. facial paralysis: see table above.

BELL'S PALSY

Definition: It is an acute paralysis of the face, due to a non-suppurative inflammation of the facial nerve near the stylomastoid foramen. It is usually unilateral, may be recurrent & sometimes runs in families.

Aetiology: Many causes have been suggested:

1. Exposure to air drafts usually precedes the onset; this may lead to ischaemia, oedema & compression of the nerve at the stylomastoid foramen.
2. It may be 2ry to a neurotropic virus e.g. Herpes zoster.
3. It may be autoimmune, as evidenced by high levels of immunoglobulins in the patient's serum.

Clinical Picture:

- The onset is usually acute with pain behind the ear.
- One or two days later, there is complete paralysis of the facial muscles on the affected side of L.M.N. nature. All 7 manifestations of L.M.N. facial lesion are seen in Bell's palsy (See p 28).
- There may be impairment of taste on the anterior $^2/_3$ of the tongue on the same side.

Treatment:

1) Medical:

1. Prednisolone tablets 30 mg/d or Synacten amp. I.M. for 2–3 weeks.
2. Vasodilators, vitamin B complex & Neostigmine.
3. Protection of the exposed cornea: eye ointment during sleep, sunglasses during daytime.

2) Physiotherapy:

1. Massage of the facial muscles.
2. Infrared irradiation to the face.
3. Galvanic stimulation to the facial muscles.

3) Surgical:

1. Decompression of the facial nerve.
2. Facial nerve grafting.
3. Plastic surgery for residual facial asymmetry.

Prognosis:

- Most patients (80%) recover in 4–6 weeks; the remaining 20% need surgical interference.
- Occasionally aberrant reinnervation occurs where the regenerating facial fibres anastomose with trigeminal fibres. Thus when the patient moves his jaw while eating or speaking there may be: – Winking (jaw–winking). – Lacrimation (crocodile tears).

8. THE COCHLEO-VESTIBULAR NERVE

This nerve is composed of 2 divisions: 1. Cochlear 2. Vestibular.

1. COCHLEAR DIVISION:

Fibres originating from the ganglion cells of the **cochlea** pass centrally from the inner ear through the internal auditory canal where they are joined by the vestibular division. They then pass through the cerebello pontine angle to enter the brain stem, where they **relay in the cochlear nucleus** (in the lower pons). In the nucleus new fibres arise; some **ascend in the** lateral lemniscus of the same side while the remainder decussate & ascend in **the lateral** lemniscus of the opposite side, to reach the **medial geniculate bodies** (M.G.B.). After **relaying** in the M.G.B. the fibres pass to the **auditory sensory area** in the temporal lobes **where** hearing is bilaterally represented.

Lesion of the cochlear division results in:

1. Tinnitus, in irritative lesions. 2. Deafness, in destructive lesions.

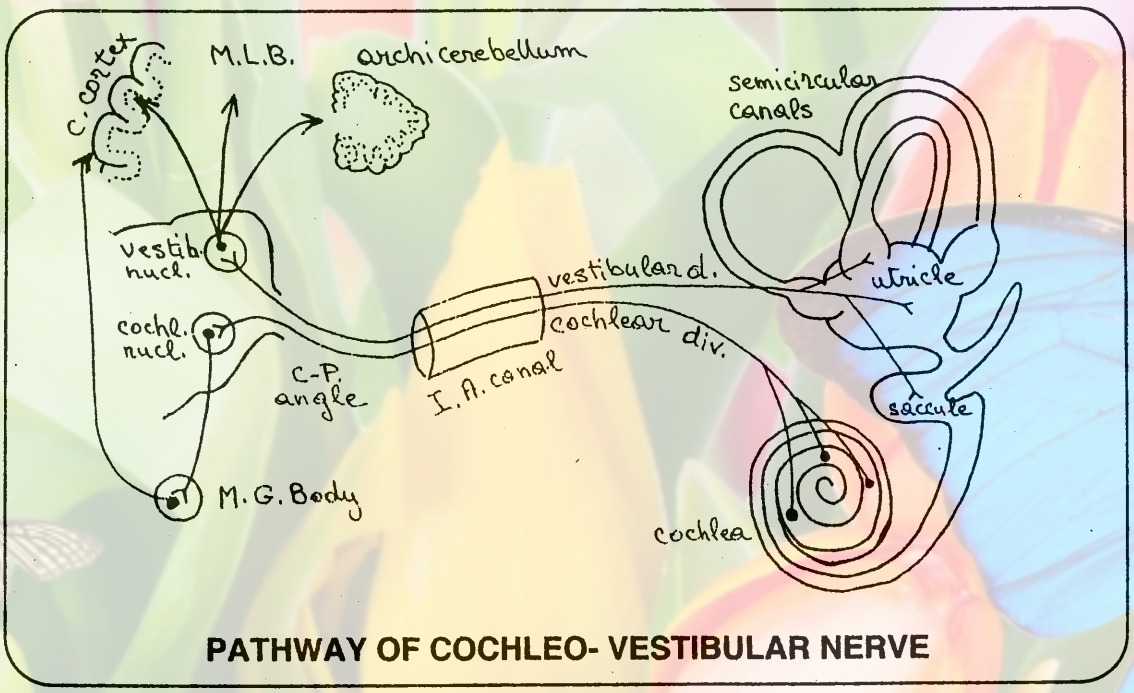
2. VESTIBULAR DIVISION:

Fibres originating from the **semicircular canal, utricle & saccule** in the inner ear **join the** cochlear division in the internal auditory canal. They then pass in the cerebello-pontine angle to enter the brain stem where they relay in the **vestibular nuclei** in the brain stem. New fibres take 3 pathways to:

1. The archicerebellum, concerned with equilibrium.
2. The medial longitudinal bundle, concerned with the synchronous movements of the eyes, head & neck.
3. The cerebral cortex, concerned with the perception of the sense of vertigo.

Lesion of the vestibular division results in:

1. Vertigo. 2. Ipsilateral incoordination. 3. Spontaneous nystagmus.



PATHWAY OF COCHLEO- VESTIBULAR NERVE

VERTIGO

DEFINITION:

- It is the sense of rotation of the body in steady surroundings or the reverse i.e.:
 - The body may be felt to rotate or fall while the surroundings are steady or
 - The surroundings themselves appear to rotate around the body.
- It is aggravated by movements of the head and it persists in all positions: sitting, standing or supine.
- It is usually associated with autonomic manifestations in the form of nausea, vomiting, pallor and bradycardia.
- Vertigo should be differentiated from dizziness, giddiness and light headedness where the unsteadiness is not associated with a sense of rotation.

CAUSES:

1) Labyrinthine:
 a) Physiological: sea sickness, car sickness.
 b) Pathological: labyrinthitis, Meniere's disease, otosclerosis.
2) Peripheral nerve:
 - Cerebello-pontine angle lesion as acoustic neuroma.
 - Vestibular neuritis.
3) Brain stem:
 - Vertebro-basilar artery insufficiency.
 - Posterior inferior cerebellar artery occlusion.
 - Disseminated sclerosis (D.S.)
 - Encephalitis.
4) Cerebral:
 - Increased intracranial tension.
 - Temporal lobe epilepsy.
5) Psychogenic:

 It occurs in acute anxiety and panic state. It responds to antidepressant and tranquilisers.

TREATMENT:

1) Treatment of the cause.
2) Cinnarizine (Stugeron), Betahistine (Betaserc) and Dramamine may be used.

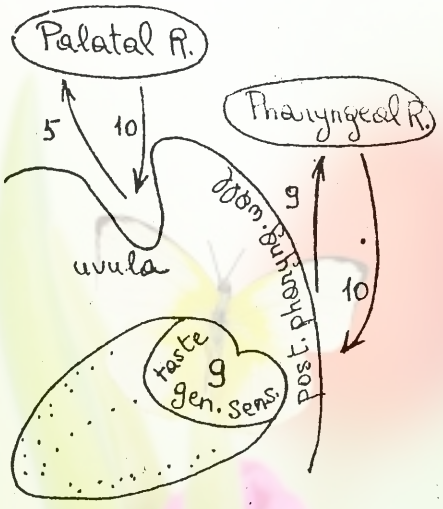
N.B.: BENIGN POSITIONAL VERTIGO:

Vertigo occurs only with a change in head position as rolling over in bed, or rising from bed or chair. It is mild and transient, lasting less than 30 seconds. It does not recur with repetition of the head movement. It is commoner in middle-aged people. Usually no treatment is required.

9. GLOSSOPHARYNGEAL NERVE

This is a mixed nerve carrying motor, sensory & autonomic (parasympathetic) fibres.

1. **Motor fibres**: to the – Stylopharyngeus.
 – Constrictors of the pharynx.
2. **Sensory fibres**:
 - General sensations from the posterior $^1/_3$ of tongue, pharynx & tonsils.
 - Taste sensation from the posterior $^1/_3$ of tongue.
3. **Autonomic fibres**:
 Parasymp. fibres to the Parotid gland.



Lesion:

1. Ipsilateral loss of taste & general sensations from the posterior $^1/_3$ of the tongue.
2. Ipsilateral loss of the pharyngeal reflex (afferent Cr 9, efferent Cr 10).

N.B.: Isolated lesions of the glossopharyngeal nerve do not occur as it is usually damaged in association with the vagus & accessory nerves at the jugular foramen (jugular foramen syndrome).

10. VAGUS NERVE

This is a mixed nerve carrying motor, sensory & autonomic (parasympathetic) fibres.

1. **Motor fibres**: to the soft palate, pharynx & larynx.
2. **Sensory fibres**: from – The skin over the external auditory meatus.
 – The thoracic & abdominal viscera.
3. **Autonomic fibres**: Parasympathetic fibres to the heart (inhibitory), the G.I.T. & the bronchial tree (secretory & motor).

Lesion:

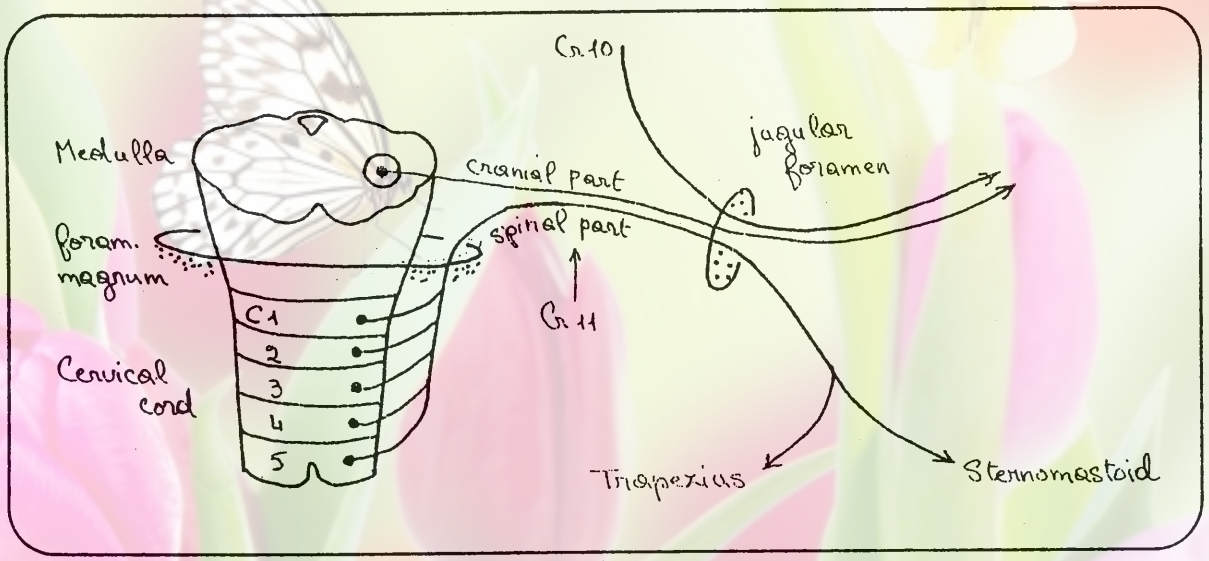
1. Palato-pharyngeo-laryngeal paralysis resulting in "**True Bulbar Palsy**" manifested by:
 - Bulbar symptoms: Dysphagia, Dysarthria, Dysphonia & Nasal regurge.
 - Ipsilateral loss of palatal & pharyngeal reflexes.
2. Tachycardia & constipation.

11. ACCESSORY NERVE

This nerve is **purely motor** & is formed of 2 parts.

1. **Cranial part**: It arises in the medulla & runs with the vagus nerve to share in the motor innervation of the soft palate & pharynx.
2. **Spinal part**: It arises from the A.H.C. of the upper five cervical segments, ascends along side the spinal cord and enters the cranial cavity through the foramen magnum. It joins the cranial portion to exit through the jugular foramen to supply the **Sternomastoid & Trapezius** muscles.

Lesion: Ipsilateral paralysis of the Sternomastoid & Trapezius muscles.



12. THE HYPOGLOSSAL NERVE

This is a **purely motor** nerve which supplies the intrinsic muscles of the **tongue**.

Lesion:

1. **U.M.N.L.:** – Unilateral: Deviation of the tongue to the opposite side of the lesion.
 – Bilateral: Inability to protrude the tongue (spastic tongue)

 In both cases there is no wasting or fasciculation.
2. **L.M.N.L.:** – Unilateral: Deviation of the tongue to the side of the lesion.
 – Bilateral: Inability to protrude the tongue.

 In both cases there is wasting and fasciculation.



Rt. L.M.N. Hypoglossal nerve lesion
(Note the wasting and deviation to the right side)

SPEECH

Development of Speech:

When the baby is born he receives sensations from the surrounding environment, which at first are meaningless, but with further development begin to acquire a meaning.

The objects seen are received as simple meaningless images in the **visual sensory area** (area 17) in the occipital lobe. Repetition of the same images leads to their insertion, storage and subsequent recognition and recall in the **visual psychic areas** (areas 18 and 19) around area 17 in the occipital lobe.

Similarly the newborn child receives auditory impulses in the **auditory sensory area** (areas 41 and 42) in the temporal lobe as meaningless sounds. With repetition of the sounds, they become inserted and stored for subsequent recognition and recall in the **auditory psychic area** (area 22) in the temporal lobe just adjacent to the auditory sensory area.

So when the child sees his mother by area 17, he recognises her in area 18, and can recall her image even in her absence in area 19. He also hears her voice in area 41 and 42 and recognises it as being hers even if he does not see her in area 22.

Later on, the child tries to imitate the sounds he has perceived. This process needs the development of a **cortical motor speech centre** known as **Broca's area** (area 44) in the frontal lobe, which is achieved by the age of 1 to 1.5 years.

When the child goes to school and starts to learn letters and numbers he begins to develop a **special visual psychic area** (area 39) in the angular gyrus in the parietal lobe for storage, recognition and recall, of their images. Area 22 also serves for the storage, recognition and recall of how they sound when spoken. When he comes to write he imitates the images of letters and numbers stored in area 39. The execution of this process of writing requires the development of a special **cortical motor centre** for writing (**Exner's area**) adjacent to Broca's area in the frontal lobe of the dominant hemisphere.

Association fibres between the visual and auditory psychic areas allow the child to correlate between the images he sees and the sounds related to them, e.g.: the sight of his mother is correlated with the word mama and vice versa. This process of correlation requires the development of an associative area (area 37) in the parietal lobe. This area also sends association fibres to Broca's and Exner's areas, and is considered as the centre in which the **ideas** of spoken or written speech is first produced.

Whether in spoken or written speech, impulses from the Broca's area or from Exner's area reach **the motor area (area 4)** descend in the pyramidal tract to reach the nuclei of the cranial nerves (supplying the muscles of articulation), and the A.H.C. (supplying the muscles concerned with writing).

Impulses from the cerebellum and extrapyramidal system share in the production of properly executed speech.

It is obvious that the process of speech involves 2 stages:

1) Speech formulation.
2) Speech articulation.

I. FORMULATION of speech, whether spoken or written is the product of 3 cooperating systems:

A) Sensory System:
 1) Visual areas: areas 17, 18, 19 and 39.
 2) Auditory areas: 41, 42 and 22.

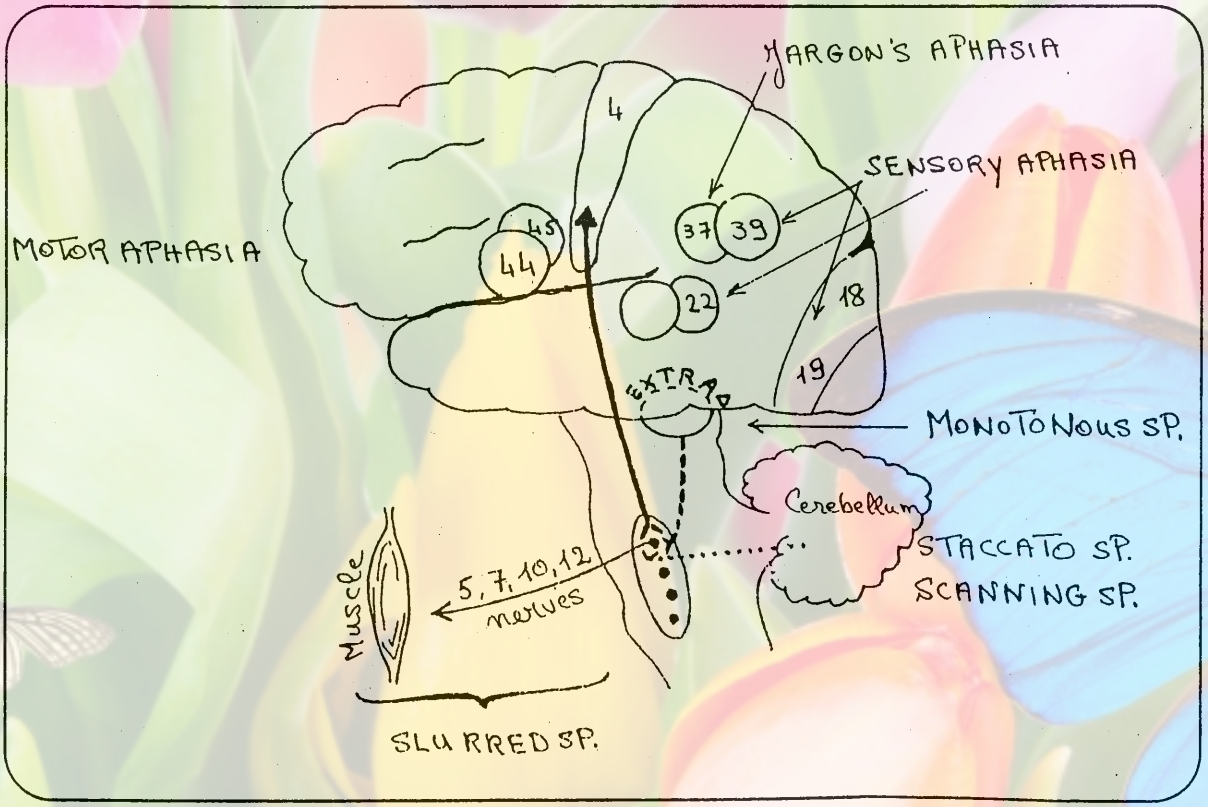
B) Motor System:
 1) Broca's area (44).
 2) Exner's area (45).

C) Associative System: area 37

N.B.: The above mentioned areas are situated in the dominant hemisphere except areas 17, 41 and 42 which are situated in both hemispheres.

II. ARTICULATION of speech is the function of the following:

1) Pyramidal tracts.
2) Cranial nuclei concerned with articulation (5, 7, 10, 12) their nerves and the muscles they supply.
3) Cerebellum for the coordination of the muscles of speech.
4) Extrapyramidal system for speech to be expressive.



APHASIA

Definition: Difficulty or inability of the formulation of speech, in the absence of lesions of the sense organs or of mental defect.

Types:

I. **Sensory or perceptive aphasia**: due to defect of perception:

1) Visual:
 - Visual agnosia: due to lesion in areas 18 and 19. The patient sees but does not recognise objects.
 - Alexia: due to lesion in area 39. The patient sees but cannot read because he does not understand the letters and numbers (word blindness).

2) Auditory:
 - Auditory agnosia: due to lesion in area 22. The patient hears but does not understand (recognise) sounds.

* Thus the patient with sensory aphasia is aphasic only in conditions, which involve the particular sensation which he cannot recognise.

II. **Motor or executive aphasia**: due to defect of execution:

1) Verbal aphasia due to lesion in Broca's area. The patient though understanding visual and auditory stimuli, yet he cannot express his ideas in spoken words.

2) Agraphia: due to lesion in Exner's area. The patient though understanding visual and auditory stimuli as well as letters and numbers, he cannot express his ideas in writing.

III. **Jargon's aphasia**: due to defect of association:

The lesion involves the associative area (area 37) and/or the associative fibres. The patient can speak, but the words are meaningless and have no relation to each other (word salad). The condition is usually associated with apraxia (for apraxia, See p. 57)

N.B.: If a child is born deaf, he will not hear sounds and therefore area 22 will not develop, hence he cannot imitate sounds, and the lack of speech in this case is termed **mutism**. Similarly if the child is born blind, he cannot see letters and numbers and hence cannot learn to write in the usual manner.

DYSARTHRIA

Definition: Difficulty in the process of articulation, though the process of formulation of speech is normal.

TYPES:

1) **Slurred speech**:

The production of consonants especially labials (e.g. B, M ...) and dentals (e.g. D, T, C...) is affected. It might be due to:

a) Bilateral pyramidal tract lesion (lesion of the cortico–bulbar fibres) as in pseudobulbar palsy.

b) Lesions involving the L.M.N. of the cranial nerves concerned with speech e.g.:
 - Nuclear: True bulbar palsy.
 - Nerve: Facial nerve palsy.
 - Neuromuscular: myasthenia.
 - Muscle: Facio–scapulo–humeral myopathy.

2) **Staccato speech**:

The speech is explosive with separation of syllables.

It occurs in cerebellar lesions as in: – Hereditary ataxias – D.S.

3) **Scanning speech** (slurred staccato): also seen in cerebellar lesions.

4) **Monotonous speech**:

The speech is expressionless and monotonous. It occurs in extrapyramidal lesions as: Parkinsonism.

N.B.: OTHER RARE TYPES OF SPEECH DISORDERS:

1) Aphonia: phonation is lost but articulation is preserved, hence the patient talks in a whisper. It is most commonly hysterical but may be organic due to disease of the larynx or of the nerve supply of the vocal cords.

2) Stuttering: Sudden interruption of the flow of speech or the repetition of words or syllables. It is usually psychogenic.

3) Palelalia: repetition of the last sentence or words of the patient's own speech. It occurs in some extrapyramidal lesions.

4) Echolalia: repetition of the last sentence or words that the patient hears. It occurs in severe dementia.

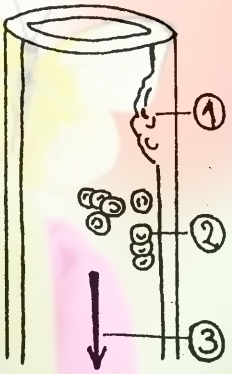
HEMIPLEGIA

DEFINITION: Paralysis of one side of the body due to pyramidal (Δ) tract lesion at any point from its origin in the cerebral cortex down to the 5th cervical segment (beginning of origin of brachial plexus).

CAUSES:

I. Vascular Causes: These are the most common.

A. Thrombotic: resulting in cerebral infarction.



1. Vessel wall diseases:
 - Cerebral atherosclerosis (most important).
 - Vasculitis e.g.: Polyarteritis nodosa & SLE.
2. Blood diseases causing hyperviscosity: they result in thrombosis.
 - Polycythemia 1ry & 2ry (↑ R.B.C.)
 - Thrombocytosis (↑ platelets)
 - Hypergammaglobulinemia (↑ gammaglobulins)
3. Circulation diseases: slow circulation results in thrombosis.
 - Heart failure.
 - Systemic hypotension (after myocardial infarction, shock, excessive hypotensive drugs, severe blood loss).

B. Embolic: resulting in cerebral infarction. The source of the embolus may be:

1. Heart (commonest):
 - Mitral stenosis with A.F.
 - Subacute bacterial endocarditis on aortic or mitral valve.
 - Mural thrombi after myocardial infarction.
 - Atrial myxoma.
 - Prosthetic valves.
 - Mitral valve prolapse.
2. Distal vessels:
 - Arterial: detached atheromatous plaque from carotid vessels or aortic arch.
 - Venous: deep venous thrombosis → detached thrombus → paradoxical embolism through a patient foramen ovale or VSD.
3. Rare sources:
 - Lungs (air emb.) in pneumothorax.
 - Tumour emboli.
 - Bones (fat emb.) in fractures.
 - Parasitic emb. (malaria, hydatid).

C. Haemorrhagic: Intracranial haemorrhage may be:

1. Intracerebral: the bleeding is in the brain substance & may leak into the ventricles; this is very serious as the blood may compress vital centres. The commonest artery causing intracerebral hge is the lenticulo–striate branch of the middle cerebral artery.
2. Subarachnoid: the bleeding is in the subarachnoid space.
3. Subdural or extradural: the blood often forms a haematoma.

THE CAUSES OF INTRACRANIAL HAEMORRHAGE ARE:

1. Hypertension: Commonest cause of intracerebral haemorrhage.
2. Rupture of an intracranial aneurysm, angioma or A–V malformation: commonest cause of subarachnoid haemorrhage.
3. Haemorrhagic blood diseases: purpura, haemophilia.
4. Anticoagulants.
5. Trauma to the head: commonest cause of subdural haematoma.

II.	**Infective:**	Encephalitis	Brain abscess.
III.	**Neoplastic:**	Meningioma	Glioma.
IV.	**Demyelinations:**	Disseminated sclerosis	Disseminated encephalomyelitis.
V.	**Traumatic:**	Cerebral laceration	Subdural haematoma.
VI.	**Congenital:**	Cerebral palsy	
VII.	**Hysterical:**	Patient suffers from paralysis in the absence of organic Δ lesion.	

CLINICAL PICTURE:

Onset & Course:

- Acute onset & regressive course (vascular, infective & traumatic lesions).
- Gradual onset & progressive course (neoplastic lesions).
- Remittent & relapsing course (D.S.).

Symptoms & Signs: Vary according to the onset:

1) Acute lesions: the clinical picture passes through 2 stages:
 1. Stage of **flaccidity** due to neuronal shock.
 2. Stage of **spasticity**; this is the stage of established hemiplegia.
2) Gradual lesions: the hemiplegia passes directly to the stage of spasticity.

I. Stage of Flaccid Paralysis (Shock Stage):

1. It lasts from 2–6 weeks, the shorter the duration the better the prognosis.
2. On the paralysed side there is complete loss of muscle tone (flaccidity), absence of deep reflexes & no plantar response.
3. This stage is prolonged by general poor condition of the patient's health, infections (e.g. chest or urinary tract infection) & bed sores.
4. During recovery from the shock stage, the muscle tone & deep reflexes reappear & gradually increase. Babinski sign becomes +ve. The stage of spasticity sets in.

5. If the onset is associated with coma, the paralysed side is determined by the following:
 - The limbs on the paralysed side are more flaccid & drop passively.
 - The cheek on the paralysed side moves in and out with respiration.

 N.B.: Don't rely on the deviation of the eyes as this may be towards the same or he opposite side of the hemiplegia, depending on the site of the lesion.

II. Stage of Spastic Paralysis stage of established hemiplegia where there is:

1. **Paralysis** of one side of the body. This paralysis shows a pyramidal distribution:
 - It affects the **progravity** more than the antigravity muscles as the former are normally weaker than the latter:

 ∴ { • In U.L. : the extensors are weaker than the flexors.
 • In L.L. : the flexors are weaker than the extensors.

 - It affects the **distal** more than the proximal muscles:

 ∴ { • The hand is weaker than the shoulder.
 • The foot is weaker than the hip.

2. **Hypertonia (spasticity)** of the paralysed muscles of clasp–knife type:
 - It affects the **antigravity** more than the progravity muscles as the former normally have the stronger muscle tone:

 ∴ { • In U.L. : the flextors are more spastic than the extensors.
 • In L.L. : the extensors are more spastic than the flextors

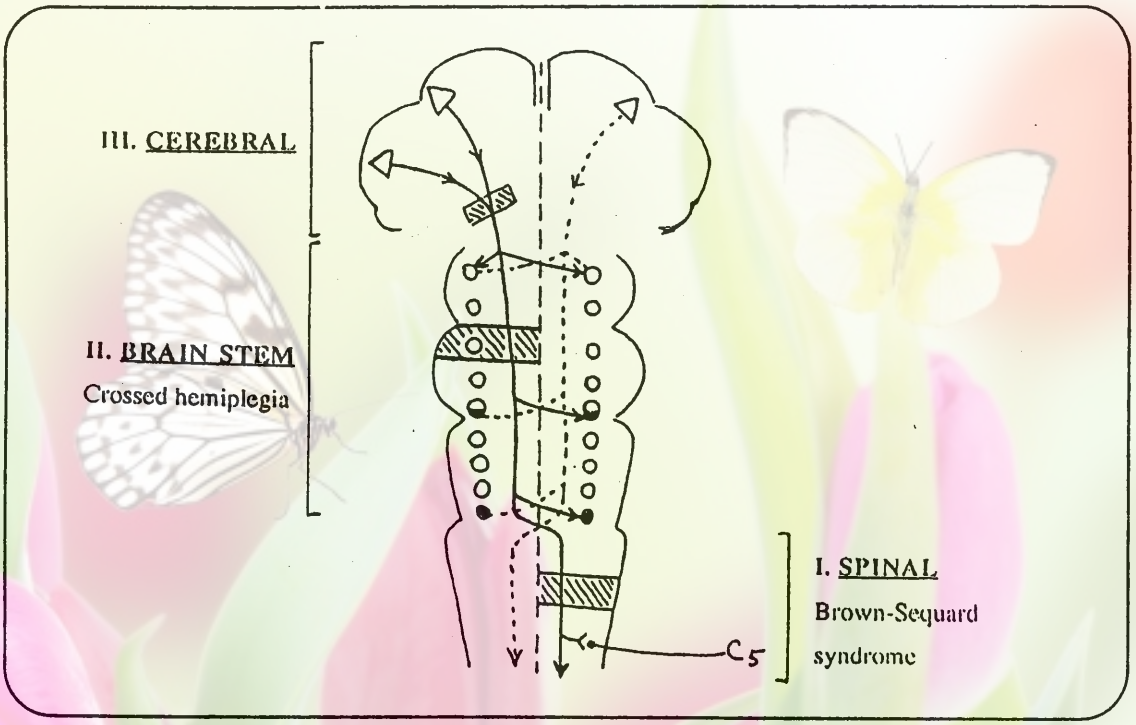
 - In both U. & L.L. **the adductors** are more spastic than the abductors.
3. **Exaggerated deep reflexes**:
 - Deep reflexes in both U. & L.L. are exaggerated on the paralysed side (biceps, triceps, brachioradialis, knee & ankle reflexes).
 - Pathological deep reflexes (normally absent) may appear:
 – finger reflex – patellar reflex – adductor reflex.
 - Clonus may be elicited in the ankle, less frequently in the knee or wrist.
4. **Lost superficial reflexes**: The abdominal and cremasteric reflexes are lost on the paralysed side.
5. **Positive Babinski sign**: On eliciting the plantar response on the paralysed side there is dorsiflexion of the big toe with or without fanning of the other toes.
6. **Gait**: If the patient can walk, his gait is circumduction due to spasticity of the extensors & adductors of L.L.

The above mentioned S & S form the classical clinical of hemiplegia; however, the C.P. may vary considerably according to:

A.The site of the lesion B. The cause of the lesion.

A. ACCORDING TO THE SITE OF THE LESION

The lesion causing hemiplegia may occur at 3 main levels:
1. Spinal cord. 2. Brain stem. 3. Cerebral.

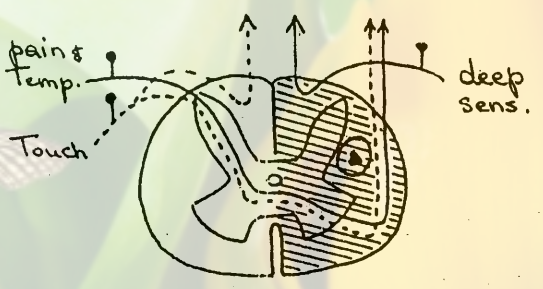


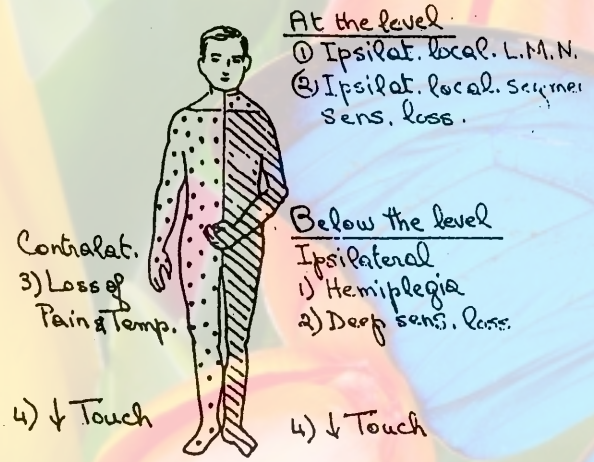
I. SPINAL CORD

The lesion is on one side of the cord & is situated between C1 & C5 segments, it is caused by: stab wound, disc prolapse, D.S. or tumour resulting in the picture of **Brown-Sequard syndrome** characterised by:

a) **At the Level of the Lesion:**
1. Ipsilateral localised L.M.N.L. of the muscles supplied by the affected segments.
2. Ipsilateral loss of all sensations in the area supplied by the dorsal roots of the affected segments.

b) **Below the Level of the Lesion:**
1. Ipsilateral hemiplegia.
2. Ipsilateral deep sensory loss.
3. Contralateral superficial sensory loss for pain & temperature.
4. Touch diminishes on both sides.





II. BRAIN STEM

The lesion is on one side of the brain stem resulting in the picture of crossed hemiplegia characterised by:

1. Hemiplegia on the opposite side of the lesion.
2. Cranial nerve paralysis of L.M.N. nature on the same side of the lesion.

The nerves affected depend on the site of the lesion:

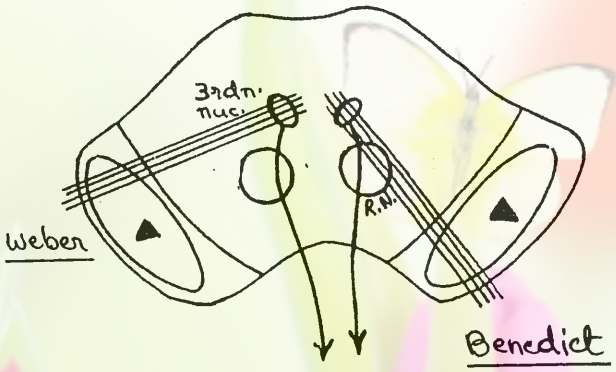
I. Mid-Brain Lesion

a) Weber's syndrome:

1. Hemiplegia on the opposite side of the lesion.
2. 3rd cranial nerve paralysis on same side of lesion.

b) Benedict's syndrome:

1. Hemiplegia on the opposite side of the lesion.
2. 3rd cranial nerve paralysis on the same side of lesion.
3. Hemiataxia (intention tremors) on the opposite side of the lesion due to affection of the Red Nucleus.



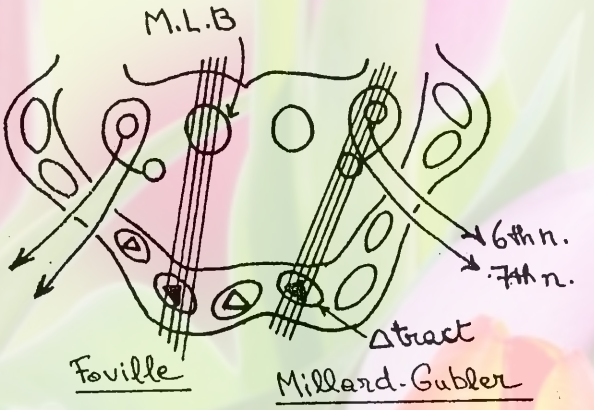
II. Pontine Lesion

a) Millard Gubler Syndrome:

1. Hemiplegia on the opposite side of the lesion.
2. 6th & 7th cranial N. paralysis on the same side of lesion.

b) Foville Syndrome:

1. Hemiplegia on the opposite side of the lesion.
2. Loss of conjugate deviation of the eyes to the same side of the lesion due to lesion in the medial longitudinal bundle (MLB)



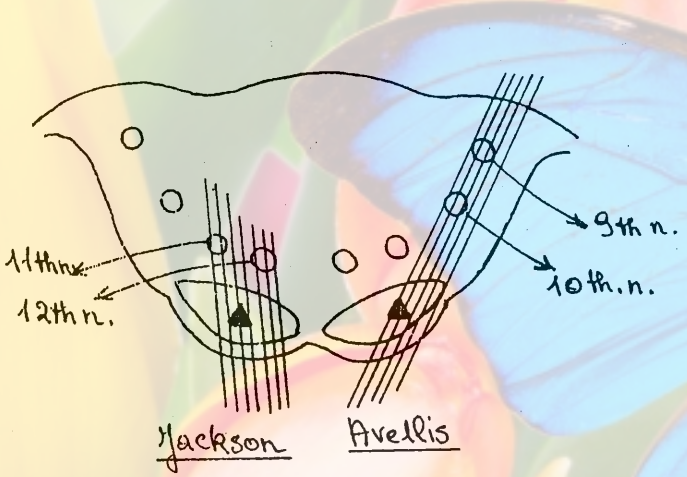
III. Medullary Lesion

a) Avellis Syndrome:

1. Hemiplegia on the opposite side of the lesion.
2. 9th & 10th cranial N. paralysis on the same side of the lesion.

b) Jackson's Syndrome

1. Hemiplegia on the opposite side of the lesion.
2. 10th & 12th cranial N. paralysis on the same side of the lesion.



III. CEREBRAL

Lesion in the cerebral hemisphere results in hemiplegia associated with U.M.N. facial and hypoglossal paralysis **on the opposite side** of the lesion, but without any cranial nerve paralysis on the same side of the lesion.

The cerebral lesion might be in one of the following sites:

Cortical: characterised by one or more of the following:

1. Coma if the lesion is extensive.
2. Convulsions if the lesion is irritative.
3. Contralateral cortical sensory loss if the parietal lobe is involved.
4. Aphasia and agraphia if the lesion is in the dominant hemisphere.
5. Homonymous hemianopia if the lesion involves the parieto–occipital region.
6. The paralysis usually involves one limb (monoplegia) specially in vascular lesions.

Subcortical: It is indistinguishable from cortical hemiplegia except that the paralysis is more extensive.

Capsular: Characterised by the following:

1. Complete hemiplegia associated with U.M.N. facial and hypoglossal paralysis on the opposite side of the lesion.
2. Hemihyposthesia on the opposite side of the lesion.
3. Hemianopia may occur, if the fibres of the optic radiation in the capsule are involved.
4. **No convulsions, aphasia or coma.**

B. ACCORDING TO THE CAUSE OF THE LESION

As the vascular causes are the commonest in hemiplegia, it is important to differentiate the clinical picture in thrombotic, embolic and haemorrhagic lesions.

Feature	Thrombosis	Embolism	Haemorrhage
1. Age	Old age	Any age	Commonly old age
2. Onset	Rapid (taking hours)	Sudden (taking sec.)	Dramatic or apoplectic
3. Prodromata	T. 1.A.S.	Absent	Absent
4. Vomiting	Absent	Absent	Common
5. Consciousness	Usually preserved	Usually preserved	Lost with deepening coma
6. Convulsions	May occur	May occur	Frequent
7. Pupils	Normal and equal	Normal and equal	Dilated and irreactive
8. Fever	Absent	Absent	Present
9. Blood pressure	May be high	Normal	Usually high
10. Heart	May be cardiac insufficiency	Usually valvular lesion*	Left ventricular hypertrophy
11. C.S.F.	Clear	Clear	Bloody, ↑ tension
12. CT scan, MRI	Hypodense area	Hypodense area	Hyperdense area

MANAGEMENT OF HEMIPLEGIA

I. GENERAL: in the acute (shock) stage of hemiplegia & in the comatosed patient:

1. Care of the skin:
 - Frequent change of the patient's position (every 2 hours), and of the bed sheets.
 - Frequent wash of the skin of the back and pressure points by alcohol followed by talc powder.
2. Care of respiration:
 - Suction of nasal and pharyngeal secretions.
 - O_2 inhalation via catheter or mask specially in cases of coma.
 - Tracheostomy in urgent cases.
3. Care of nutrition and fluid balance:
 Tube feeding giving fruit juices, milk and pureed food, besides I.V. fluids, in comatosed patients.
4. Care of the urinary bladder:
 - Foley's self–retaining catheter in case of retention or incontinence.
 - Urinary antiseptics: Gentamycin 80 mg I.M. every 12 hours.
5. Care of the bowels: Daily enema.

II. SYMPTOMATIC:

1. Cerebral dehydrating measures (Mannitol, Glycerol or Steroids) to minimise vasogenic brain oedema. However, repeated injections of Mannitol is avoided as it may lead to a severe rise of blood pressure or to renal damage.
2. Antiemetics in suppository form (Largactil or Primperan) and potassium in cases of severe vomiting.
3. Tranquilizers and sedatives (Paraldehyde) in cases of insomnia and irritability.
4. Muscle relaxants (Sirdalud or Coltramyl) for spasticity.
5. Vitamin and tonics.

III. PHYSIOTHERAPY:

1. Proper positioning of the hemiplegic side.
2. Massage, passive and active exercises (after stabilisation of the neurological condition) to minimise contractures & to strengthen the muscles.

IV. SPECIFIC: It is the treatment of the cause.

A) Cerebral Thrombosis:

1) **Care of Blood Pressure**:
 a. Hypotensive drugs if the blood pressure is above 200/120 e.g.:
 Captopril (Capoten). Dose: 25–50 mg t.d.s.
 Avoid sudden marked reduction of B.P. which decreases cerebral blood flow and worsens the brain ischaemia.
 b. Vasopressor drugs if the blood pressure is very low (as in myocardial infarction).
2) **Antiplatelets**: because platelet aggregation is increased after thrombosis.
 1. Aspirin: single dose 75–300 mg/daily, or
 2. Persantin (Dipyridamole): 75 mg twice daily, or
 3. Ticlid (Ticlopidine): 250 mg twice daily.
3) **Anticoagulants**: They are not used in all cases.
 Indications:
 1. Stroke in evolution i.e. gradual progressive weakness (over days) denoting gradual & progressive thrombus formation.
 2. Recurrent T.I.A.s although they are less effective in preventing stroke than antiplatelet drugs.
 3. Embolic hemiplegia specially in cardiac cases to prevent recurrent embolisation.

 Contraindications:
 1. Haemorrhagic infarction; it is excluded by using the CT scan.
 2. Haemorrhagic blood disease.
 3. Subacute bacterial endocarditis (SBE).
 4. Excessive salicylate intake
 5. Renal & hepatic failure.
 6. High fevers.

 Method: start by Heparin 5000 U/6 hours I.V. (immediate action) together with oral anticoagulants Dindivan or Marcoumar: 150 mg (3 tab) 1st day – 100 mg (2 tab) 2nd day – 50 mg (1 tab) 3rd day (action starts after 48 hrs). After 48 hrs stop heparin & continue oral anticoagulants. Monitor the dose using the prothrombin time.

 * ***Antidote***: – Heparin: protamine sulphate – Oral anticoagulants: vitamin K.
4) **Other drugs may be used:**
 1. **Nootropil (Piracetam)**: It improves brain metabolism by increasing oxygen consumption; it also decreases blood viscosity by reducing platelet aggregation.
 Dose: 12 gm/day I.M. or I.V. in acute cases.
 2. **Trental (Pentoxifylline)**: It improves the microcirculation of the brain by increasing RBC deformability & reducing platelet aggregation.
 Dose: 800–1200 mg/day infusion in acute cases.

3. **Trivastal (Piribedil)**: It is a dopamine agonist which increases cerebral blood flow by its antivasoconstrictive effect. Dose: 20 mg t.d.s. orally.
4. **Cerebral vasodilators**: as CO_2 in oxygen (carbogen), papaverine . . . etc. are less commonly used nowadays because overall cerebral vasodilatation may divert blood from the ischaemic area (**steal phenomenon**).

B) Cerebral Embolism:

1. **Treatment of the source of emboli**.
2. **Anticoagulants** are given in cardiac cases to prevent further embolisation.
 Method: See before.

C) Cerebral Haemorrhage:

1. **General care** (see p. 45):
 1. Care of the skin.
 2. Care of respiration.
 3. Care of nutrition.
 4. Care of the urinary bladder.
 5. Care of the bowels.
2. **Hypotensive drugs** in cases of hypertension.
3. **Antifibrinolytic drugs** as tranexamic acid & amino caproic acid. They delay clot lysis & thus prevent rebleeding but may increase cerebral ischaemia.
4. **Coagulants** as vitamin K are no longer used.
5. **Surgical** evacuation of the haematoma provided it is not intraventricular.

D) Cerebral Inflammation:

1. Meningitis: sulphadiazine.
2. Encephalitis: corticosteroids.

E) Brain Tumours:

1. Surgical removal.
2. Deep x–ray therapy.

CAUSES OF TRANSIENT HEMIPLEGIA

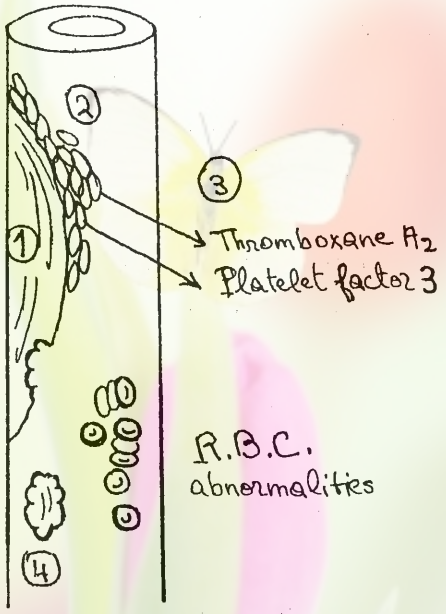
1. Transient ischaemic attacks TIAs.
2. Hypertensive encephalopathy.
3. Hemiplegic migraine.
4. Cerebral aneurysm.
5. Demyelinating disease e.g. DS.
6. Post–epileptic Todd's paralysis.
7. Congestive attacks of GPI in syphillis.
8. Hysterical.

PATHOGENESIS OF CEREBROVASCULAR (CV) INSUFFICIENCY

1) Cerebral Atherosclerosis:

It is the commonest cause of CV insufficiency leading to thrombotic lesions of the brain.

1. Atheromatous plaques narrow the lumen of the vessel.
2. The rough surface of the plaques allow platelet–aggregation with further reduction and prevention of blood flow.
3. The release of several substances from the clump of platelets (thromboxane A_2, platelet factor 3 and others . . .) leads to:
 a) Vasoconstriction.
 b) Activation of blood coagulation (fibrinogen → fibrin) resulting in clot formation.
4. Fragments of atheroma and thrombi pass into the circulation and act as emboli.



2) RBC abnormalities Normal RBC are highly deformable to allow them to pass easily through small capillaries; while rigid cells retain their shape and cannot pass through the microcirculation; they aggregate, increase blood viscosity causing CV insufficiency and thrombotic lesions of the brain.

3) Risk factors predisposing to cerebro–vascular insufficiency e.g. hypertension, diabetes, hyperlipidaemia, hyperuricaemia, obesity, chronic stress, smoking . . .

Males are more commonly affected.

* These above factors lead to a reduction of cerebral blood flow and ischaemia of the brain cells.

If the ischaemia lasts a short period its clinical manifestations will be transient (e.g. TIA).

If the ischaemia lasts longer it will result in cerebral infarction which may be:

1. **A large infarction:** if the vessel occluded is a major artery or a large branch.
2. **A small (lacunar) infarction:** if the vessel occluded is one of the small perforating arteries.

TRANSIENT ISCHAEMIC ATTACKS (T.I.A.S)

DEFINITION: These are attacks of cerebral ischaemia too brief to cause infarction and usually lasting few minutes or hours (max. 24 hours) from which the patient recovers completely.
If it lasts over 24 hours and then the patient recovers, it is termed Reversible Ischaemic Neurological Deficit (RIND)

CAUSE: 1. Microemboli arising from atheromatous plaques in the large cerebral vessels (carotid or vertebro–basilar) or from the heart. This the commonest cause.
2. Vasculitis (e.g. SLE, polyarteritis nodosa) and hyperviscosity states (polycythaemia, sickle–cell anaemia) are less common causes.

CLINICAL PICTURE: The patients present with the manifestations of either:

1. Carotid insufficiency; any one of the following: transient hemiparesis, aphasia, convulsions, unilateral blindness (amaurosis fugax) ... etc.
2. Vertebro–basilar insufficiency; any one of the following: transient diplopia, vertigo, unsteadiness, circumoral numbness . .. etc.

INVESTIGATIONS:

1. Doppler ultrasonic imaging: It is used on the neck vessels and it maps out an image of the moving column of blood in the vessel.
2. Real time (B mode) ultrasound imaging: shows a longitudinal or transverse section of the vessel.
3. Digital subtraction angiography: It visualises the intracerebral vessels. It is non–invasive as it requires a contrast medium given I.V.
4. Cerebral angiography: It is invasive and requires a contrast medium injected directly into the artery via a catheter. It is the most precise method, showing any occlusion or stenosis in the cerebral vascular tree.
5. Investigations for the risk factors as: blood glucose, serum uric acid, serum lipogram..

PROGNOSIS: Patients suffering from T.I.A.s are more liable to develop cerebral strokes.

TREATMENT:

A. Medical:

1) **Antiplatelet aggregating drugs**: they reduce the incidence of strokes by about 50%.
 1. Acetyl–salicylic acid: 75–300 mg daily.
 2. Dipyridamole (Persantin): 75 mg twice daily.
 3. Ticlopidine (Ticlid): 250 mg twice daily is very effective but has many side effects.
2) **Anticoagulant drugs** as Dindivan or Marcomar; are less effective in preventing strokes.
3) **Other drugs** used with the antiplatelet drugs as Nootropil, Trental or Trivastal.
4) **Treatment of any risk factor**

B. Surgical:

1) Endarterectomy is used in carotid artery stenosis of over 70% to relieve recurrent TIAs and to prevent a major stroke. It is not used in mild stenosis or if a stroke has already occurred.
2) Angioplasty of distal carotid or middle cerebral artery is still being tested.

LACUNAR INFARCTIONS:

These are small infarcts (0.5–1.5 cm in diameter) produced by occlusion of the penetrating branches of the major cerebral arteries and are mainly due to sustained hypertension. These infarcts are termed "lacunes" as they are filled with fluid and they are found in the internal capsule, thalamus, basal ganglia and pons.

CLINICAL PICTURE:

1. Lacunar infarcts may be asymptomatic or
2. They may present with: pure motor hemiplegia (lesion in the capsule), pure sensory stroke (lesion in the thalamus), ataxia (lesion in the pons) ... depending on the site of the lesion.
3. C.T. scan: shows small hypodense areas at the sites of the lacunes.

TREATMENT: control of the hypertension.

HYPERTENSIVE ENCEPHALOPATHY:

This is an acute commonly **reversible** disorder of cerebral function due to severe i.e. malignant hypertension (diast. B.P. above 140 mmHg). Normally, a rise in B.P. causes a reactive vasoconstriction of the cerebral arteries to protect the brain against flooding with blood. In hypertensive encephalopathy there is failure of this autoregulation leading to severe cerebral vasodilatation and clinical manifestations of encephalopathy.

CLINICAL PICTURE:

1. Encephalopathic manifestations: headache, vomiting, blurred vision, confusion, drowsiness and maybe coma; convulsions may occur.
2. Focal manifestations: blindness, aphasia, hemiparesis.
3. Fundus examination: bilateral papilloedema, exudates and hypertensive retinopathy.

PROGNOSIS: The condition is reversible as usually there is no infarction.

TREATMENT:

- Hypotensive drugs produce dramatic relief.
 Sodium nitroprusside (Nipride): 1–5 μg/kg/min infusion.
- Anticonvulsant drugs e.g. Valium or Phenytoin may be needed in cases of convulsions.

BLOOD SUPPLY OF THE BRAIN

The blood reaches the brain through two systems of blood vessels:

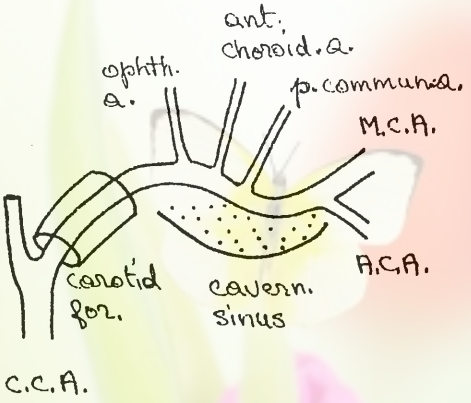
1. The carotid system. 2. The vertebral system.

I. THE CAROTID SYSTEM:

Each internal carotid artery enters the cranial cavity through the carotid foramen and canal to the cavernous sinus where it lies lateral to the optic chiasma.

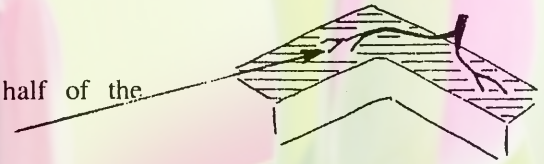
The artery in the sinus gives off three small branches:

- The ophthalmic artery.
- The anterior choroidal artery.
- The posterior communicating artery.



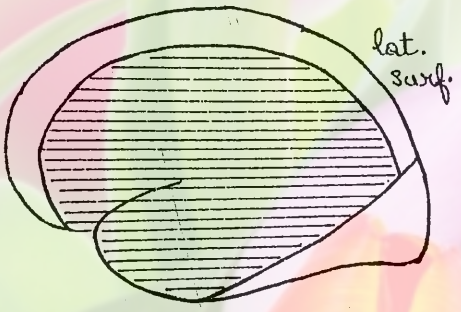
The internal carotid artery then divides into its two main terminal branches the middle and anterior cerebral arteries.

1) **The middle cerebral artery** runs on the lateral surface of the cerebral hemisphere, in the lateral (Sylvian) sulcus to supply the lateral aspect of the anterior $^3/_5$ of the cerebral hemisphere.



* It gives the following branches:

1. **Capsular** branch supplying the dorsal half of the internal capsule (Lenticulo–striate artery).
2. **Cortical** branches:
 a) *Frontal branch* supplying the lower part of the motor area (face, U.L. and trunk), and the motor speech areas.
 b) *Parietal branch* supplying the lower part of the sensory area, the angular and supramarginal and the upper fibres of the optic radiation.
 c) *Temporal branches* supplying the auditory areas and the lower fibres of the optic radiation.

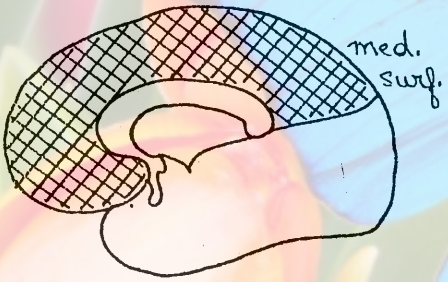


2) **The anterior cerebral artery** runs medially on the medial surface of the cerebral hemisphere, then around the corpus callosum as far as the parieto–occipital sulcus. This artery supplies the medial aspect of the anterior $^3/_5$ of the cerebral hemisphere and the upper edge of the lateral surface.



* It gives the following branches:

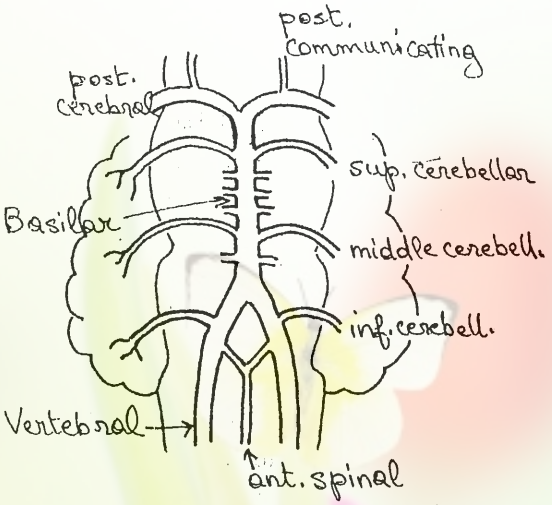
1. **Capsular** branch (Heubner's artery) supplying the ventral half of the anterior limb of the internal capsule.
2. **Cortical** branches:
 a) *Frontal branch* supplying the pre–frontal area (area of mentality and inhibition of primitive reflexes).
 b) *Paracentral branch* supplying the motor and sensory areas of the L.L., and the paracentral lobule (cortical bladder centre).
 c) *Callosal branch* supplying the corpus callosum.



II. THE VERTEBRAL SYSTEM:

Each vertebral artery passes upwards through the vertebral foramina to enter the cranial cavity through the foramen magnum and runs upwards on each side of the medulla. Both arteries meet at the lower border of the pons to form one midline single artery, the basilar artery, which runs upwards on the ventral surface of the pons were it gives small branches known as the paramedian arteries to the brain stem and divides into its two terminal branches the posterior cerebral arteries.

Each posterior cerebral artery supplies the whole occipital lobe and the posterior part of the temporal lobe (posterior $^2/_5$ of the cerebral hemisphere).



VERTEBRO-BASILAR SYSTEM

* It gives the following branches:

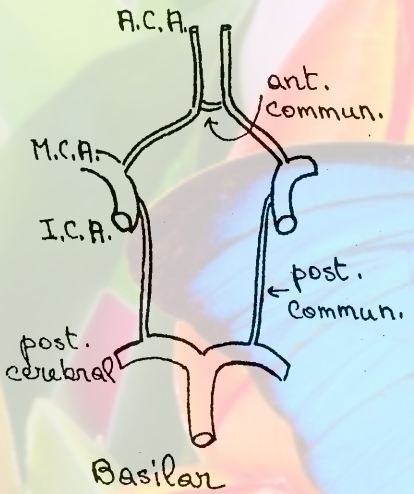
1) **Posterior communicating** artery.
2) **Capsular** branch supplying the ventral half of the posterior limb of the internal capsule, the thalamus and the geniculate bodies (Thalamogeniculate artery).
3) **Cortical** branches to the occipital lobe.

In its course the vertebro–basilar system gives:

- Two **spinal arteries** which unit to form the anterior spinal artery.
- Three **cerebellar arteries** on each side: The superior middle and inferior cerebellar arteries.

CIRCLE OF WILLIS:

Both anterior cerebral arteries are connected together by the anterior communicating artery. Also, the internal carotid artery of each side is connected to the posterior cerebral artery of the same side by the posterior communicating artery. In this way the circle of Willis is formed where the two carotid arteries communicate with each other and with the vertebro–basilar system.



CIRCLE OF WILLIS

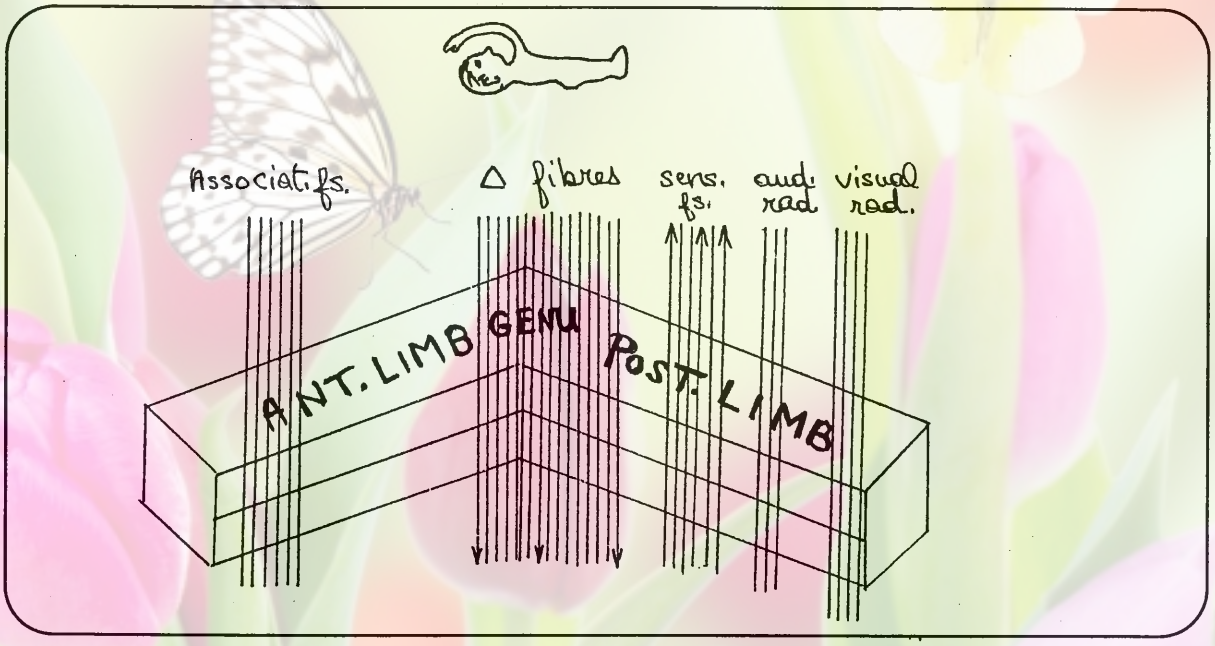
INTERNAL CAPSULE I.C. :

It is a broad band of white fibres lying in the depth of the cerebral hemisphere. It is formed of:

1. The anterior limb A.L. placed between the caudate nucleus medially and the lentiform nucleus laterally.
2. The genu.
3. The posterior limb P.L. placed between the thalamus medially and the lentiform nucleus laterally.

Fibres passing through the I.C.:

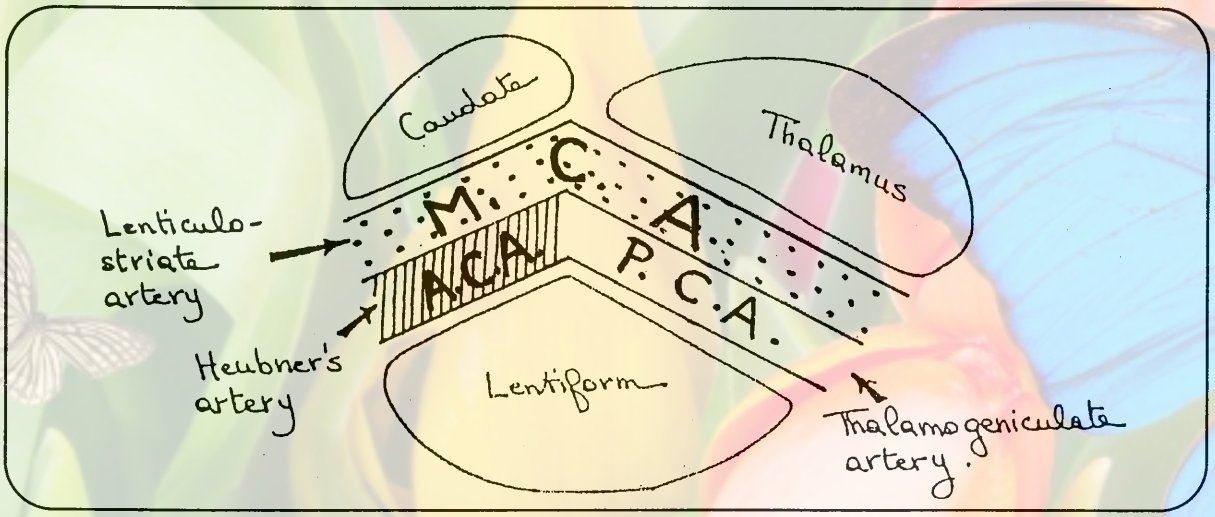
1. Δ fibres descend in the genu, adjacent part of AL and ant. $^1/_2$ of PL. The fibres supplying the arm are followed by those supplying the head, trunk, and lastly the lower limb.
2. Sensory fibres from the thalamus ascend in the PL.
3. Auditory and optic radiations pass in the posterior part of PL.
4. Associative fibres pass in the anterior part of AL.



BLOOD SUPPLY:

The **dorsal half** of the anterior limb, knee, and posterior limb of the internal capsule is supplied by the capsular branches of the middle cerebral artery (Lenticulo striate artery).

The **ventral half** of the anterior limb of the I.C. is supplied by the capsular branches of the anterior cerebral artery (Heubner's artery). The ventral half of the knee and posterior limb is supplied by the capsular branches of the posterior cerebral artery (thalamo–geniculate artery).



VASCULAR OCCLUSIVE SYNDROMES

INTERNAL CAROTID ARTERY OCCLUSION

- Age: any age may be affected but most commonly the 4th –6th decades.
- Sex: males are more affected than females.
- Aetiology: the commonest is atherosclerosis of the artery.
- Onset: it may be preceded by recurrent transient attacks of one or more of the following:
 1. Headache.
 2. Blindness (Amaurosis fugax).
 3. Hemiparesis or hemihyposthesia.
 4. Aphasia.
 5. Convulsions.
 6. Mentality changes.
- These symptoms indicate internal carotid artery insufficiency (**carotid T.I.A.s**) which may terminate in complete occlusion of the artery manifested by:
 1. Ipsilateral blindness.
 2. Contralateral hemiplegia.
 3. Contralateral hemihyposthesia.
 4. Contralateral homonymous hemianopia.
 5. Aphasia with or without agraphia in left–sided lesions.
 6. The internal carotid pulse may be diminished or lost and an audible bruit over the vessel may be heard.

MIDDLE CEREBRAL ARTERY OCCLUSION

I. Main Artery Occlusion:

1. Coma at the onset.
2. Contralateral hemiplegia affecting U.L. more than L.L.
3. Contralateral hemihyposthesia with cortical sensory loss in U.L.
4. Contralateral homonymous hemianopia.
5. Aphasia and agraphia in left sided lesions.

II. Capsular Branch Occlusion: (Lenticulo striate artery)

1. Contralateral complete hemiplegia affecting the upper and lower limbs to the same extent.
2. Contralateral hemihyposthesia of subcortical type.
3. Contralateral hemianopia may occur.
4. No loss of consciousness or aphasia.

III. Cortical Branches Occlusion:

a) Frontal vessel occlusion:

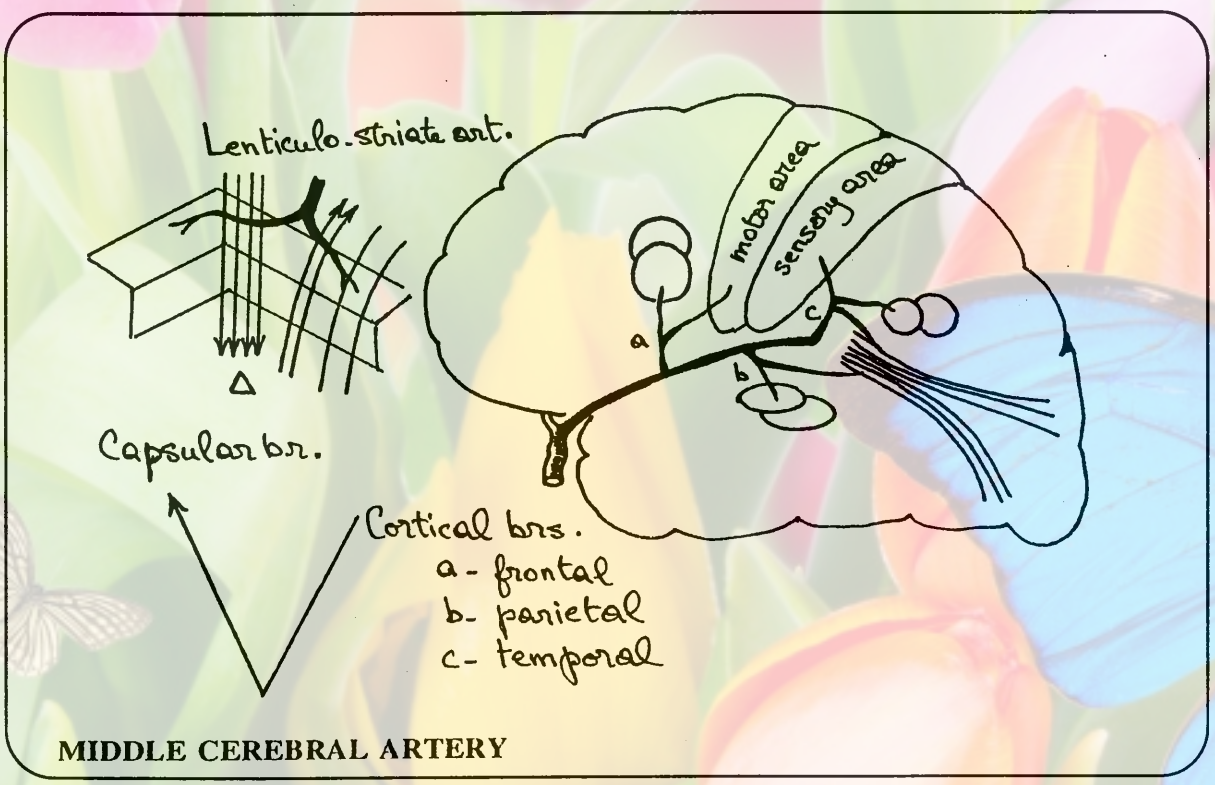
1. Facio–brachial monoplegia.
2. Motor aphasia and agraphia in left–sided lesions.

b) Parietal vessel occlusion:

1. Cortical sensory loss in the upper limb.
2. Lower quadrantic homonymous hemianopia.
3. Sensory aphasia (alexia) and apraxia in left sided lesions.

c) Temporal vessel occlusion:

1. Upper quadrantic homonymous hemianopia.
2. Sensory aphasia (auditory agnosia).



MIDDLE CEREBRAL ARTERY

ANTERIOR CEREBRAL ARTERY OCCLUSION

I. Main Artery Occlusion:

1. Contralateral hemiplegia affecting L.L. more than U.L.
2. Contralateral cortical sensory loss in the lower limb.
3. Incontinence of urine.
4. Mentality and personality changes.
5. Forced grasp reflex.

II. Capsular Branch Occlusion (Heubner Artery):

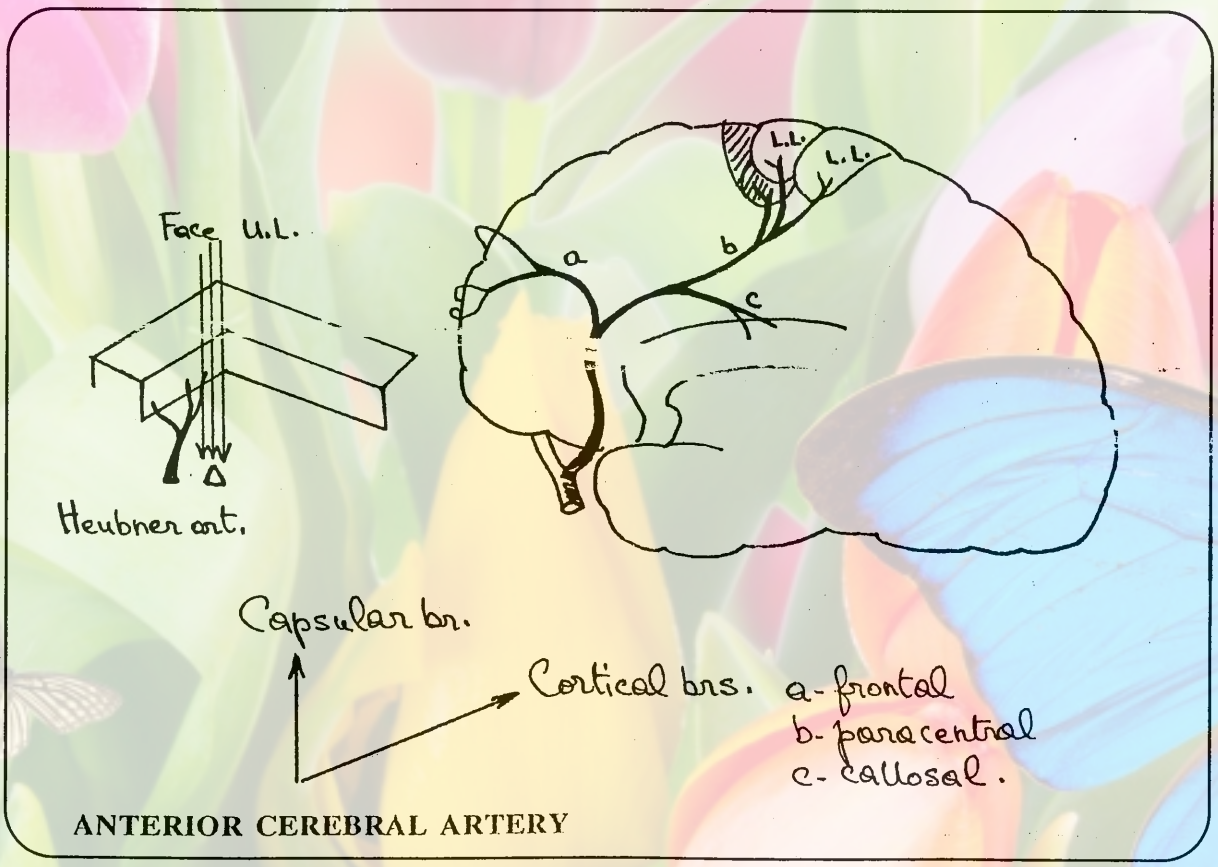
- Facio–brachial monoplegia which is proximal more than distal, i.e. involving the shoulder more than the hand.

III. Cortical Branches Occlusion:

a) Frontal vessel occlusion:
 1. Mentality and personality changes.
 2. Forced grasp reflex.

b) Paracentral vessel occlusion:
 1. Monoplegia in the lower limb.
 2. Cortical sensory loss in the lower limb.
 3. Urinary incontinence.

c) Callosal vessel occlusion:
 - Apraxia of the left side.



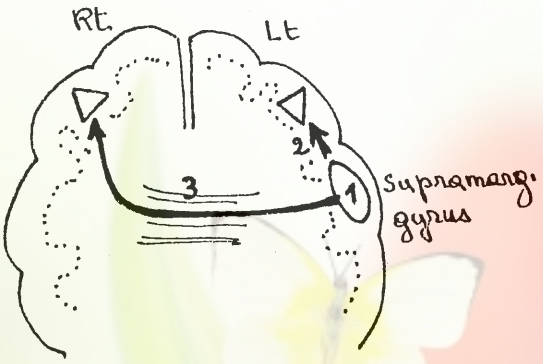
ANTERIOR CEREBRAL ARTERY

APRAXIA:

It is the inability to perform compound voluntary motor activity in the absence of paralysis, sensory loss or incoordination.

It might be due to a lesion at:

1. The supramarginal gyrus of the dominant hemisphere resulting in apraxia of both sides
2. The fibres connecting the supramarginal gyrus to the motor area of the dominant hemisphere resulting in apraxia of the right side.
3. The anterior part of the corpus callosum containing the fibres from the supramarginal gyrus of the dominant hemisphere to the non–dominant one resulting in apraxia of the left side.



POSTERIOR CEREBRAL ARTERY OCCLUSION

I. Main Vessel Occlusion:

1. Contralateral homonymous hemianopia with macular sparing as the macula has double blood supply.
2. Visual agnosia in left sided lesions.
3. Thalamic syndrome; in cases where the occlusion is proximal to the origin of the thalamogeniculate artery.

II. Ganglionic Branch: (Thalamogeniculate artery) Occlusion: Thalamic Syndrome:

1. Thalamic pain i.e. constant burning severe pain in the hemianaesthetic side.
2. Complete contralateral hemianaesthesia to all types of sensations with partial recovery of the superficial sensations; however the vibration sense is permanently lost.
3. Reflex dystrophy of the shoulder girdle and arm, secondary to the pain.
4. Choreo–athetosis or hemiballismus due to ischaemia of the basal ganglia and/or subthalamus.

III. Cortical Branches Occlusion:

1. Contralateral homonymous hemianopia with macular sparing.
2. Visual agnosia in left sided lesions.

VERTEBRO-BASILAR ARTERY OCCLUSION

I. Main Vessel Occlusion:

a) Partial occlusion (insufficiency): manifested by recurrent transient attacks of the following (**V. Basilar T.I.A.s**).
 1. Syncope.
 2. Diplopia.
 3. Ophthalmoplegia.
 4. Vertigo or tinnitus.
 5. Bulbar symptoms (dysphagia, dysarthria, nasal regurgitation, and hoarseness of voice).
 6. Hemiparesis, hemianaesthesia or paresthesias.
 7. Ataxia.

b) Complete occlusion (usually fatal):
 - Deep coma.
 - Complete quadriplegia with decerebrate rigidity.
 - Bulbar paralysis.
 - Respiratory embarrassment.

II. Cerebellar Arteries Occlusion:

a) Superior cerebellar artery occlusion:
 1. Ipsilateral cerebellar ataxia.
 2. Ipsilateral Horner's syndrome.
 3. Ipsilateral deafness.
 4. Contralateral hemihyposthesia.

b) Middle cerebellar (anterior inferior cerebellar) artery occlusion:
 1. Ipsilateral cerebellar ataxia.
 2. Ipsilateral Horner's syndrome.
 3. Ipsilateral deafness.
 4. Ipsilateral 5th, 6th and 7th cranial nerve paralysis.
 5. Contralateral hemihyposthesia

c) Inferior cerebellar **(posterior inferior cerebellar)** artery occlusion:

This artery arises as a branch of the vertebral artery but may rarely originate from the proximal part of the basilar artery. It runs upwards on the lateral aspect of the medulla supplying its upper lateral part which contains:

1. Some of the reticular formation nuclei.
2. Part of the vestibular nucleus.
3. Spinal sensory nucleus of the 5th nerve for pain and temperature sensations of the same side of the face.
4. Sympathetic fibres to the eye and face.
5. The nuclei of 9th, 10th and 11th cranial nerves.
6. Spinal lemniscus carrying pain and temperature sensations from the opposite side of the body.

The artery finally terminates and supplies the inferior part of the cerebellum.

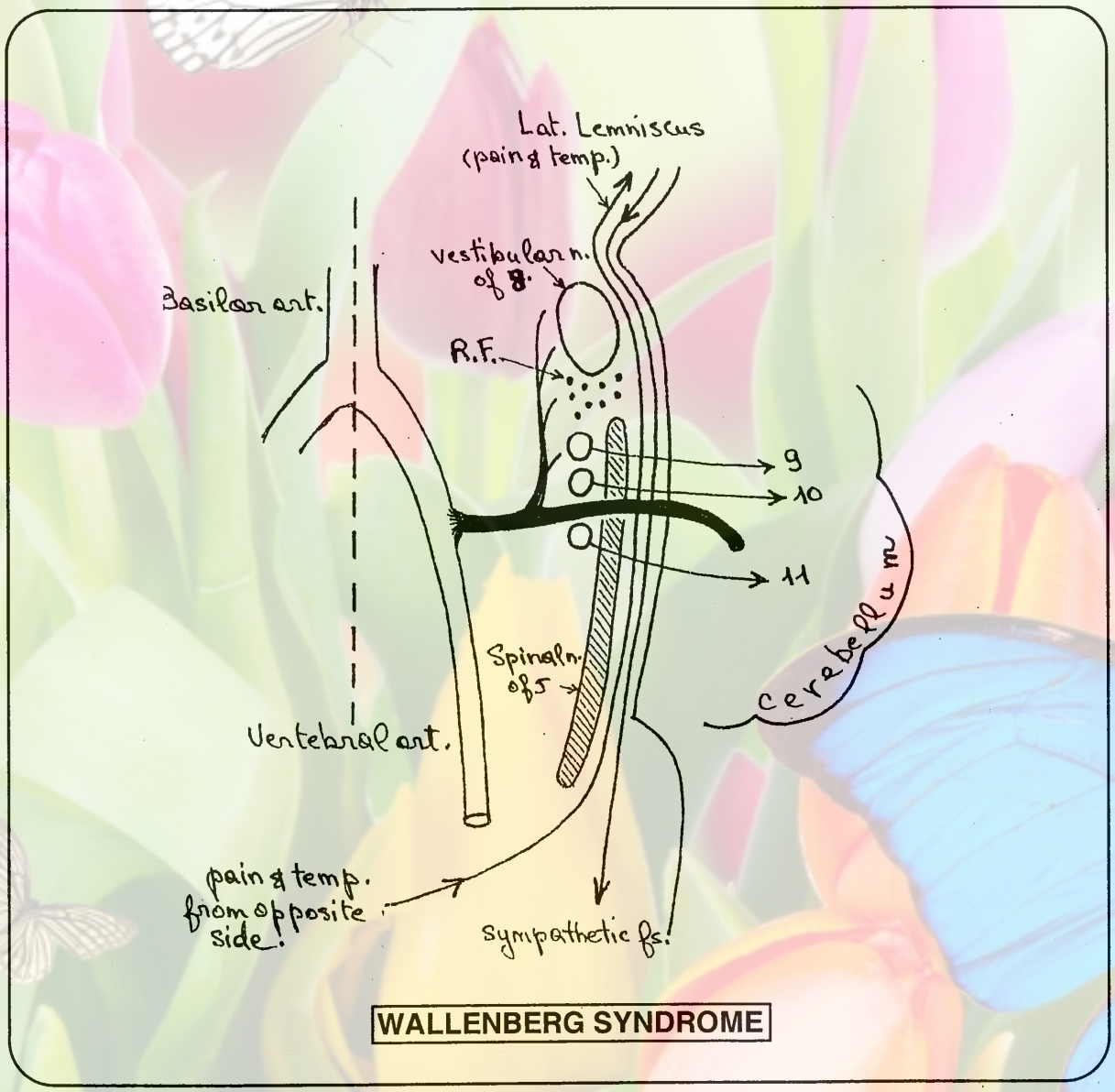
Occlusion of the artery will lead to **Wallenbergs syndrome:**

1. Acute onset associated with syncope, hiccup, vomiting, vertigo and pain over the face.
2. Ipsilateral cerebellar ataxia (nystagmus, dysarthria, incoordination).
3. Ipsilateral Horner's syndrome.
4. Ipsilateral palato–pharyngeo–laryngeal paralysis and weakness of the sternomastoid and trapezius.
5. Ipsilateral loss of pain and temperature sensations over the face.
6. Contralateral loss of pain and temperature sensations over the body.

III. Brain Stem Branches Occlusion:

a) Mid–brain: – Weber's syndrome. – Benedict's syndrome.

b) Pons: – Millard–Gubler's syndrome. – Foville's syndrome.



WALLENBERG SYNDROME

PARAPLEGIA

Paraplegia is paralysis or weakness (paraparesis) of both lower limbs. It may be due to:

- Pyramidal (U.M.N.) lesion resulting in **spastic** paraplegia.
- Lower motor neurone (L.M.N.) lesion resulting in **flaccid** paraplegia.

SPASTIC PARAPLEGIA

DEFINITION:

It is paralysis or weakness of both lower–limbs due to bilateral pyramidal tract lesion, most commonly in the spinal cord (spinal paraplegia), and less commonly in the brain stem or the cerebral parasagittal region (cerebral paraplegia).

Spinal paraplegia may be:
1. Focal: paraplegia **with a level**.
2. Systemic
3. Disseminated

} paraplegia **without a level**

CAUSES OF SPINAL PARAPLEGIA:

(I) Focal Causes:

A. Compression:

1. Vertebral:.
 - Fracture or fracture–dislocation of the vertebra.
 - Disc prolapse and spondylosis.
 - Pott's disease.
 - Neoplastic diseases:
 - Primary as osteosarcoma or haemangioma.
 - Metastatic from a 1ry carcinoma of the thyroid, breast, lung, stomach, kidney & prostate.
 - Deformity of the vertebral column as kyphoscoliosis.
2. Meningeal (extramedullary):
 - Extradural e.g., leukaemic deposits.
 - Dural e.g., meningioma, pachymeningitis hypertrophica of syphilis.
 - Intradural e.g., neurofibroma, cystic arachnoiditis.
3. Cord (intramedullary):
 – Syringomyelia – Glioma or Ependymoma of the cord.

B. Inflammatory:

– Transverse myelitis – Myelomeningitis – Myeloradiculitis.

C. Vascular: Anterior spinal artery occlusion.

II.Systemic Causes:

A systemic disease in neurology is a disease which affects one or more systems selectively and is usually bilateral and symmetrical. When a systemic disease affects the pyramidal tracts, either alone or with other tracts, paraplegia will result.

A. Heridofamilial:

1. Hereditary spastic paraplegia.
2. Hereditary ataxias e.g., Friedreich's or Marie's ataxia.

B. Symptomatic:

1. Pellagral lateral sclerosis.
2. Subacute combined degeneration.

C. Idiopathic: motor neurone disease.

III.Disseminated Causes:

A disseminated disease in neurology is a multifocal disease of the same nature:

1. Disseminated Sclerosis (D.S.)
2. Disseminated Encephalo–myelitis (D.E.M.)
3. Disseminated Syphilitic lesions.

CAUSES OF CEREBRAL PARAPLEGIA:

A. Causes in the Parasagittal Region: (area of cortical presentation of L.L.)

1. Traumatic e.g.
 - Depressed fracture of the vault of the skull.
 - Subdural haematoma.
2. Vascular e.g., superior sagittal sinus thrombosis.
3. Inflammatory e.g., encephalitis, meningio–encephalitis.
4. Neoplastic e.g., parasagittal meningioma.
5. Degenerative e.g., cerebral palsy (Little's disease).

B. Causes In The Brain Stem:

- Syringobulbia.
- Midline brain stem tumours.

These lesions arise in the midline and involve the innermost pyramidal fibres which are those of the lower limbs.

CLINICAL PICTURE OF FOCAL PARAPLEGIA

A. AT THE LEVEL OF THE LESION:

1. Vertebral manifestations: only present if the cause is vertebral.

– Localised pain or tenderness. – Localised deformity or swelling.

2. Radicular manifestations: only present in extra–medullary causes.

a) Posterior root affection:

- Early pain in the back referred to the distribution of the affected root and described as girdle pain; it is exaggerated by coughing, sneezing and straining.
- Later there is hyposthesia or anaesthesia in the dermatome supplied by the affected root.

b) Anterior root affection: localised L.M.N. weakness in the muscles supplied by the affected root.

N.B.: As the lesion in paraplegia is below the cervical segments (which supply the muscles of the arm), the L.M.N. affection at the level of the lesion is not clinically evident; this is due to the difficulty in eliciting wasting and the difficulty of testing for tone and deep reflexes in the trunk and abdominal muscles. If the lesion involves the cervical segments there is quadriplegia with evident signs of L.M.N.L. in the upper limbs.

B. BELOW THE LEVEL OF THE LESION (CORD MANIFESTATIONS):

1. MOTOR MANIFESTATIONS: They depend on whether the cause of the lesion is acute or gradual.

a) **If the cause is acute** (inflam., vascular or traumatic). The paraplegia passes by 2 stages:

- Stage of **flaccidity** due to **neuronal shock**:

 Immediately following the lesion there is sudden paralysis of the lower limbs, associated with complete loss of tone and absence of reflexes (**flaccid paralysis**). This stage lasts from 2 to 6 weeks.

- Stage of **spasticity** due to recovery from the neuronal shock:

 On recovery from the shock stage, the fullblown picture of U.M.N.L. will be established including: hypertonia, hyperreflexia, positive Babinski sign & maybe clonus.

b) **If the cause is gradual** (e.g., neoplastic):

The shock stage is absent, and there will be gradual progressive weakness of the lower limbs with hypertonia and hyperreflexia:

1. The *weakness* affects the distal group of muscles more than the proximal group and the flexor muscles more than the extensors.
2. The *hypertonia* and *hyperreflexia* affect the extensor group of muscles (antigravity) more than the flexor group (prograviry) The paraplegia in this stage is described as (**paraplegia in extension**)
3. With further progression of the lesion, the extrapyramidal fibres in the cord will be affected. The hypertonia and hyperreflexia will be more in the flexor group of muscles than in the extensors. In this stage the paraplegia is described as (**paraplegia in flexion**).

 This last stage may be associated with the **mass reflex** where there is spontaneous urination, defaecation and sweating on scratching the **skin over the** medial side of the thigh associated with reflex erection and ejaculation on squeezing the glans penis.

N.B.: Piere Marie Foix test is done by firm passive plantar flexing of the toes and **foot. This** will result in spontaneous "withdrawal reflex" i.e. spontaneous flexion of the hip, **knee and** dorsiflexion of the ankle if the paraplegia is passing from extension to flexion.

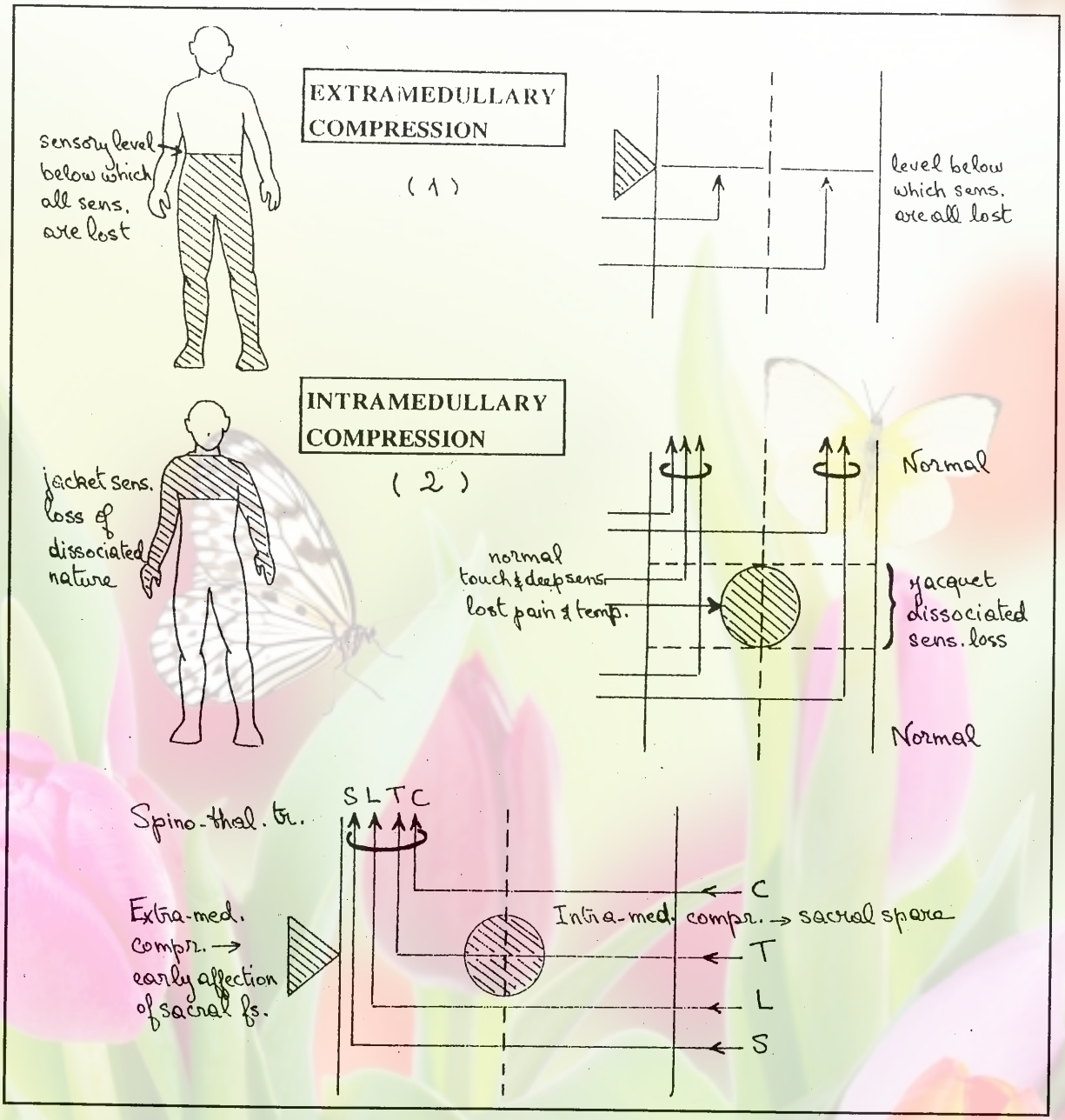
	Paraplegia in extension	Paraplegia in flexion
1. Cause	Pyramidal lesion	Pyramidal and extrapyramidal
2. Hypertonia	More in extensors	More in flexors
3. Position of L.L.	Extended	Flexed
4. Deep reflexes	Exaggerated	Less exaggerated
5. Clonus	Present	Absent
6. Mass reflex	Absent	May be present
7. Bladder	Precipitancy	Automatic bladder

2. SENSORY MANIFESTATIONS:

a) If the cause of the lesion is **extramedullary**, encroachment on the ascending tracts at the site of lesion results in **a sensory level** below which all types of sensations are diminished. There is early loss of sensation in the saddle area (S 3, 4, 5), as the sacral fibres lie in the outermost part of the spinothalamic tracts in the cord.

b) If the cause of the lesion is **intramedullary**, there will be a **jacket sensory loss** (hyposthetic area with normal sensations above and below it). The sensory loss is of a **dissociated nature** i.e., pain and temp. sensations are lost but touch and deep sensations are preserved; this is due to the interruption of the crossing fibres carrying pain and temp. by the midline lesion, while touch and deep sensation fibres ascend in the posterior column without decussation.

The sensations over the saddle area are preserved (**sacral spare**), as the sacral fibres lie far from the midline lesion.

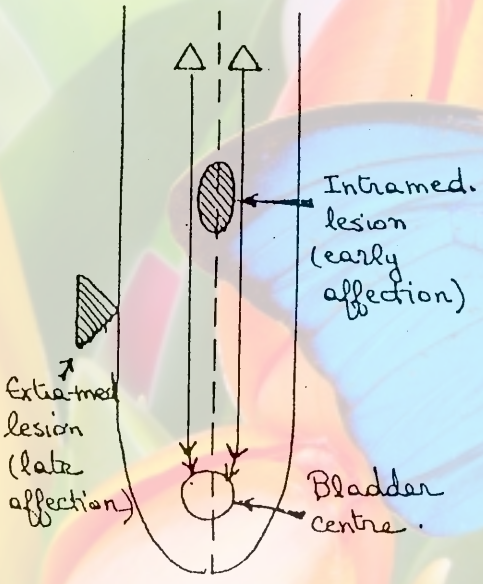


3.SPHINCTERIC MANIFESTATIONS:

a) In acute lesions retention of urine in the shock stage, followed by precipitancy of micturition.

b) In gradual lesions: precipitancy of micturition which may terminate in automatic bladder when complete transection of the cord occurs.

* These changes start late in extramedullary lesions and early in intramedullary lesions as the pyramidal fibres controlling the bladder centre lie medially in the cord.

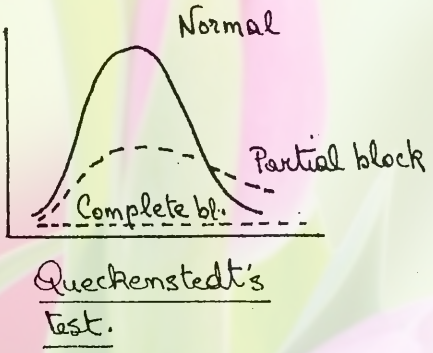


INVESTIGATIONS:

1) C.S.F. Examination

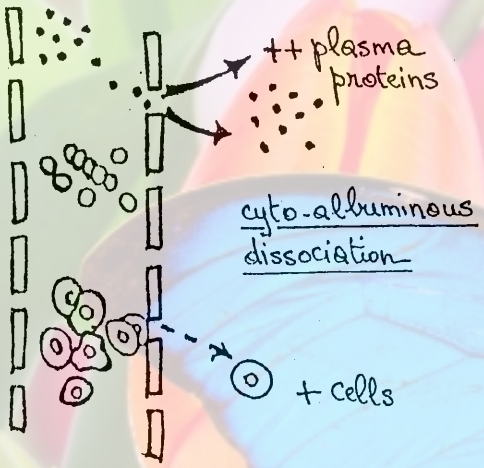
Normally the C.S.F.; is a clear colourless fluid, formed by the choroid plexus in the cerebral ventricles. It leaves the ventricles through the lateral and medial foramina of the 4th ventricle to the subarachnoid space where it is absorbed by the subarachnoid villi to the intracranial venous sinuses to be finally drained into the internal jugular veins.

1. **C.S.F. pressure**: it is measured by tapping the C.S.F. in the lumbar region, using a needle connected to a spinal manometer.
 - Normally it is 120 ml of C.S.F. or water.
 - In extramedullary compression it is markedly diminished.
 - In intramedullary compression it is less markedly diminished.
2. **Quecknestedt's test**:
 - Bilateral jugular vein compression below the angle of the mandible normally leads to rapid rise of C.S.F. pressure in the manometer; release of the compression leads to rapid drop of pressure to normal.
 - In extramed. compr. there is no change of C.S.F. pressure (**complete dynamic block**) due to complete obstruction of the subarachnoid space.
 - In intramed. compr. there is a slower and lesser rise of pressure (**partial dynamic block**) due to incomplete obstruction.



Queckenstedt's test.

3. **C.S.F. constituents:**
 - Normally:
 - Proteins 20–40 mg/100 ml.
 - Cells 0–5/ H.P.F. mainly lymphocytes.
 - Chlorides 720–750 mg/100 ml.
 - Sugar 50–80 mg/100 ml.
 - In extramed. compr. there is marked increase in proteins leading to spontaneous coagulation and yellowish discolouration (**xanthochromia**), the cell count is rather normal (**cyto–albuminous dissociation**).
 - In intramed. compr. there is no marked change



cyto-albuminous dissociation

N.B.: The association of xanthochromia, cyto–albuminous dissociation and spontaneous coagulation of the C.S. is known as **Froin's syndrome**.

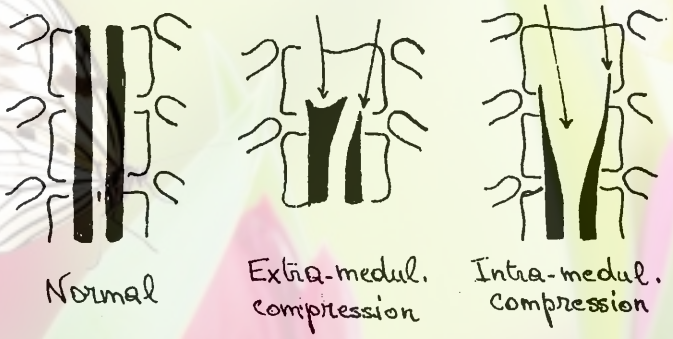
2) Plain X–Ray of the Spine: It may show:

- Destruction of the vertebrae as in Pott's disease and metastases.
- Narrowing of the intervertebral spaces with or without osteophytes as in spondylosis and disc prolapse.
- Fracture or dislocation of the vertebrae.

3) Myelography:

A water soluble radio–opaque substance (metrizamide) is injected into the subarachnoid space and its movement is viewed on a screen.

- Normally: the dye moves freely throughout the subarachnoid. space.
- In extramed. compr. there is complete block with short tails (saddle shape).
- In intramed. compr. there is increased width of the cord with long tails (fusiform shape).



	Extra-medullary	Intra-medullary
I. History:		
1. Duration	– Long	– Short
2. Onset	– Painful due to posterior root irritation	– Painless
3. Bladder disturbances	– Absent or late	– Early
II. Clinical picture:		
1. Motor	– Usually asymmetrical	– Usually symmetrical
2. Sensory	– Sensory level.	– Jacket sensory loss.
	– All types of sensations are diminished below the level including pain, temp. and touch.	– Dissociated sensory loss i.e., loss of pain and temp. with preservation of touch.
	– Early loss of sensation in the saddle area.	– Late loss of sensation in the saddle area (sacral spare)
3. Sphincteric	– Late bladder disturbances.	– Early bladder disturbances.
III. Investigations:		
1. C.S.F.	– Marked lowering of pressure.	– Less marked lowering of pressure.
	– Complete dynamic block.	– Partial or no dynamic block.
	– Froin's syndrome: spontaneous coagulation, xanthochromia, cyto–albuminous dissociation	– No spontaneous coagulation, xanthochromia, or cyto–albuminous dissociation
2. Plain x–ray	– Possible vertebral lesion.	– Normal x–ray.
3. Myelography	– Saddle shaped block.	– Fusiform shape.

MANAGEMENT OF PARAPLEGIA:

I. General: (this is mainly the nurse's job).

- Frequent change of the patient's posture to guard against bedsores and hypostatic pneumonia.
- Care of the skin by frequent washing with alcohol followed by talc powder. In case of urinary incontinence frequent change of the bed–sheets.
- Care of the bladder: if there is retention use parasympathomimetic drugs as prostigmine (1–2 ampoules I.M./6 hours). If this fails use a catheter to evacuate the bladder.

II. Physiotherapy:

1. Massage to increase the blood supply to the paralysed muscles.
2. Passive exercises to guard against fibrosis and stiffness.
3. Active exercises to strengthen the muscles.
4. Positioning: The paralysed limb is put in a position slightly opposite to the hypertonia.

III. Symptomatic Treatment:

1. Analgesics and sedatives for pain.
2. Muscle relaxants e.g., myanesin or coltramyl or valium for the spasticity.
3. Vitamins and tonics.

IV. Specific Treatment: (treatment of the cause) e.g.:

1. Antituberculous drugs in case of Pott's disease.
2. Deep X–ray in case of intra–medullary tumour.
3. Surgical eradication in case of extra–medullary tumour.

TRANSVERSE MYELITIS

Definition: It is an acute lesion involving the gray and white matter of a limited number of spinal cord segments.

Aetiology:

1) Infection: – Viral: Influenza, chicken pox, post–vaccination.
- Bacterial: T.B., Syphilis, diphtheria.

2) Toxic: Post lumbar anaesthesia. 3) Demyelinating; D.S., and D.E.M.

Clinical Picture:

- Shock stage (2–6 weeks): Flaccid paraplegia with complete loss of sensation below the level of the lesion and retention of urine.
- Recovery stage: Spastic weakness of the lower limbs with hypothesia below the level of the lesion and precipitancy of micturition.

Investigations: C.S.F. examination: increased cells and proteins, in D.S. there is high immunoglobulin level.

ANTERIOR SPINAL ARTERY OCCLUSION

It may be caused by thrombosis, embolism or a dissecting aortic aneurysm.
Its clinical picture is the same as that of transverse myelitis but there is **sparing of the deep sensations** as the posterior columns of the spinal cord are supplied by the posterior spinal artery.

SYRINGOMYELIA

Definition: It is a chronic disease characterised by gliosis and cavitation around the central canal in the gray matter of the spinal cord specially in the lower cervical and upper thoracic segments and to a lesser extent in the lumbar segments.

Pathology: The disease is due to the proliferation of congenital undifferentiated cell rests of glial tissue in the gray matter of the spinal cord. The excess glial tissue being avascular will undergo central necrosis and cavitation with destruction of the surrounding structures.

Clinical Picture:

– Age: Commonly between 15 and 35 years.

– Onset and course: Gradual onset and slowly progressive course.

– S. and S.: These depend on whether the syringomyelia is cervical or lumbar.

a) **Cervical syringomyelia**:

1. Motor manifestations:
 – ***Early***: In the U.L.: Localised L.M.N. weakness with fasciculations due to encroachment on A.H.C.
 – ***Late***: In the L.L.:Spastic paraplegia due to encroachment on the pyramidal tracts.
2. Sensory manifestations: **Jacket sensory** loss of **dissociated** nature in the area of skin supplied by the affected segments with sacral spare.
3. Autonomic manifestations: Trophic ulcers of the fingers, painless nail infection and vasomotor changes (**Morvan's syndrome**).
4. Associated skeletal anomalies as pes cavus, and spina bifida are common.

b) **Lumbar syringomyelia**: Picture of epiconus lesion.

Investigations: Myelography is diagnostic.

Treatment:

1. X–ray irradiation of the affected region of the cord.
2. Physiotherapy and symptomatic treatment.

SYRINGOBULBIA

– It is a medullary lesion similar to that in syringomyelia.

– The lesion may start in the medulla or may be the upward extension of a cervical syringomyelia.

– It involves the spinal nucleus of Cr. 5, the vestibular and bulbar nuclei.

– **Clinically** it presents with:

- Pain in the face followed by loss of sensation of dissociated nature.
- Vertigo.
- Bulbar symptoms with lost palatal and pharyngeal reflexes.
- Wasting of the tongue.

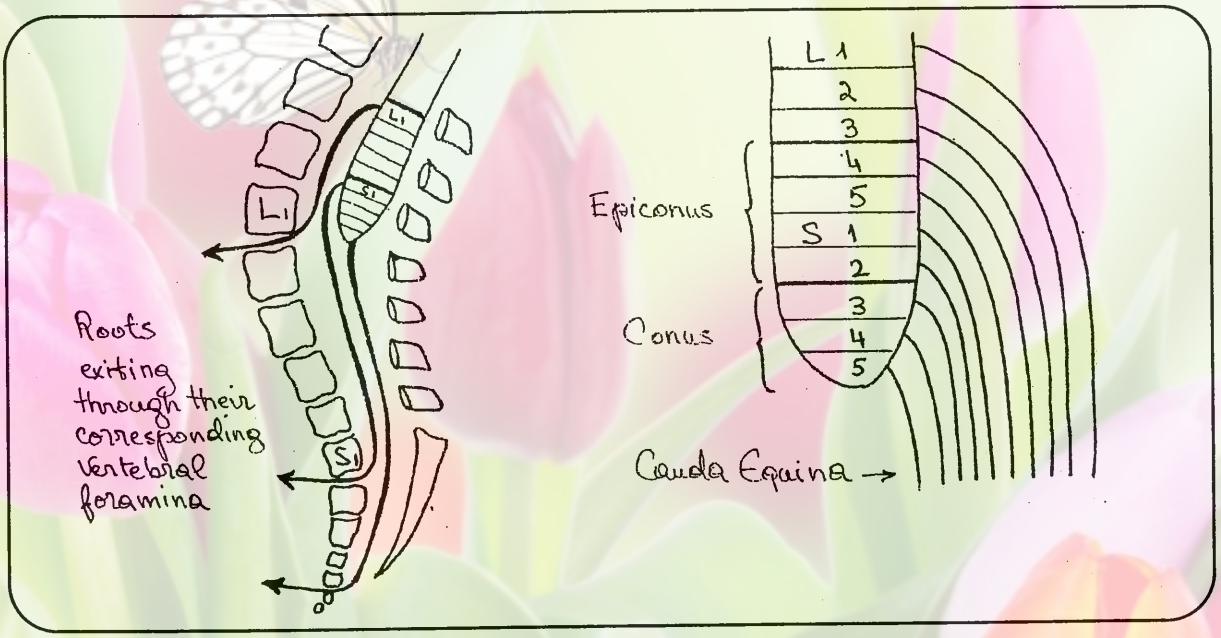
CAUDA EQUINA

ANATOMY:

During intra–uterine life the rate of growth of bones is faster than the rate of growth of the soft tissue so that, at birth, the vertebral column (bones) is longer than the spinal cord (soft tissue). Normally the lower–most end of the spinal cord is at the level of the lower border of the first lumbar vertebra or the upper border of the second lumbar vertebra (i.e. at the junction between the first and second lumbar vertebrae). From this level downwards, the spinal canal is not empty, it is filled by the collection of the lumbo–sacral roots which descend in this space to escape through their corresponding intervertebral foramine.

This collection of lumbo–sacral roots in the lower part of the spinal canal is known anatomically as the **Cauda Equina**. The lowermost three segments of the spinal cord ($S_{3,4,5}$) are known anatomically as the **Conus Medullaris**. The above four segments ($L_{4,5}$ $S_{1,2}$) are known anatomically as the **Epiconus**.

N.B.: The cauda equina consists of nerve roots while the conus and epiconus form part of the spinal cord.



CAUSES OF CAUDA EQUINA LESIONS:

1. Congenital: Spina bifida.
2. Traumatic:
 – Fracture or fracture dislocation of the lumbar vertebrae.
 – Post traumatic disc prolapse.
3. Inflammatory: Pott's disease of the lumbar vertebrae.
4. Neoplastic:
 a) Vertebral:
 – Primary: osteoma, haemangioma.
 – Secondary: i.e., metastatic.
 b) Meningeal: meningioma.
 c) Radicular: neurofibroma.
5. Degenerative: Lumbar Spondylosis.

CLINICAL PICTURE OF CAUDA EQUINA LESIONS:

Cauda equina lesions may present by one or more of the following manifestations:

I. Motor Manifestation:

- There is motor weakness or paralysis in one or both lower limbs.
- The weakness or paralysis is of a L.M.N. nature i.e. it is associated with wasting, hypotonia and hyporeflexia.
- The motor weakness or paralysis will affect the muscles which are supplied by the affected root. The function of each root can be easily tested in the following muscles:

Root	Action	Muscles
L_2	Flexor of the hip	Ileopsoas.
L_3	Extensor of the knee	Quadriceps
L_4	Dorsiflexion of the ankle	Anterior tibial group
L_5	Dorsiflexion of the toes	Anterior tibial group & glutei
S_1	Plantar flexion of the ankle and toes	Calf muscles & glutei
S_2	Flexor of the knee	Hamstrings
$S_{3, 4, 5}$	Anal contraction	Anal and perianal muscles

II. Sensory Manifestations:

- Cauda equina lesions usually have a painful onset. The pain is radicular and is referred to the lower limbs, either along the femoral distribution when the lesion affects the upper lumbar roots or along the sciatic distribution when the lesion affects the lower lumbar. and sacral roots. Later on there is hyposthesia or anaesthesia in the dermatome supplied by the affected root.
- The sensory impairment affects both superficial and deep sensations.

Root	Sensory
L_1	Upper third of the front of the thigh.
L_2	Middle third of the front of the thigh
L_3	Lower third of the front of the thigh.
L_4	Antero–lateral aspect of the thigh, Front of the knee, Antero–medial aspect of the leg, Medial aspect of the dorsum of the foot and big toe.
L_5	Lateral aspect of the thigh and leg, Middle third of the dorsum of the foot and Middle three toes.
S_1	Postero–lateral aspect of the thigh and leg, Lateral third of dorsum of the foot and little toe.
S_2	Posterior aspect of the thigh and leg and sole of the foot.
$S_{3, 4, 5}$	Anal, perianal and gluteal region (saddle–shaped area).

III. Autonomic Manifestations:

a) Sphincteric manifestations are usually late unless the lesion is bilateral and affects mainly $S_{2,3,4}$ roots (roots of innervation of the bladder). The sphincteric disturbances are in the form of:.

1. Sensory atonic bladder. 2. Motor atonic bladder or 3. Autonomic bladder.

b) Vasomotor changes and trophic ulcers may occur in the L.L.

CLINICAL PICTURE OF CONUS MEDULLARIS LESION: (S3,4,5 SEGMENT) :

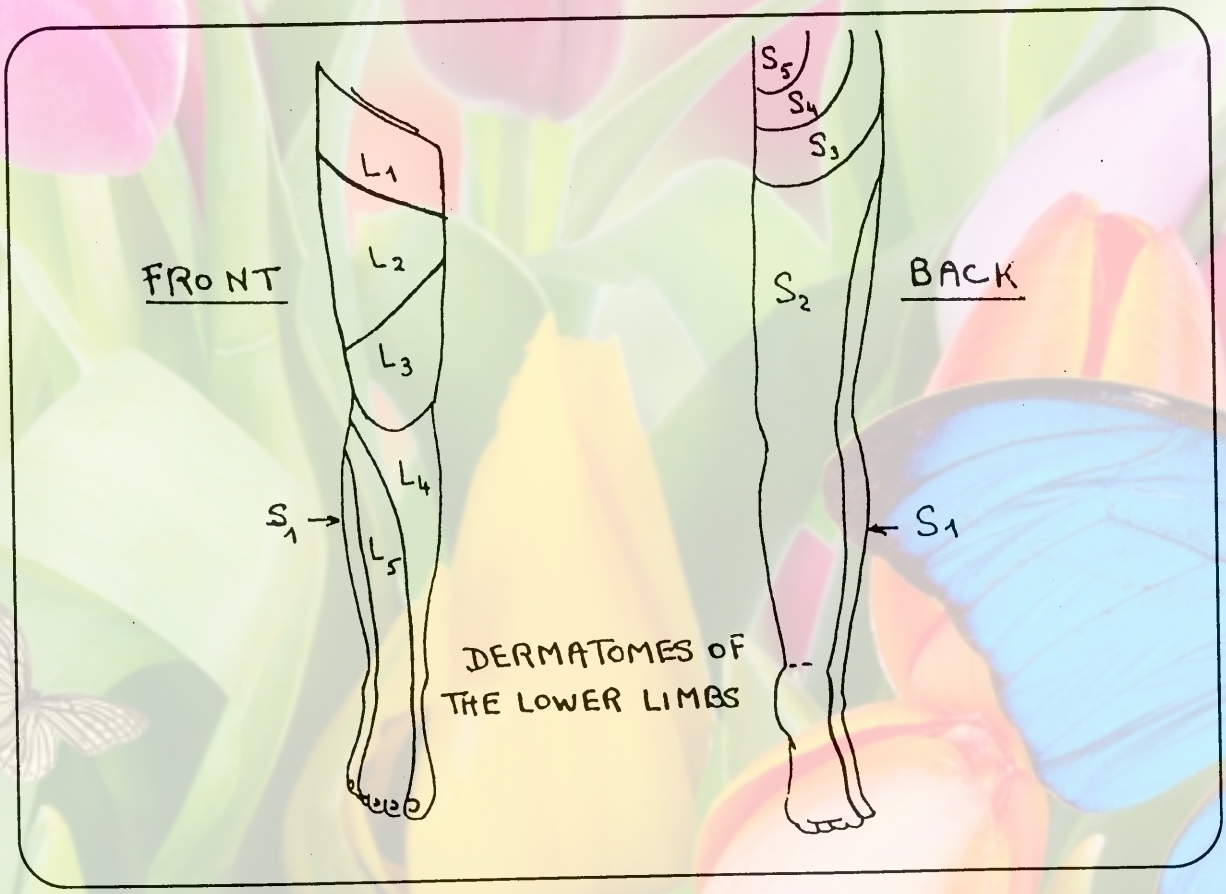
1. Early urinary incontinence (autonomic bladder) and faecal incontinence.
2. Impotence.
3. Impairment of sensation in the saddle–shaped area, (usually of a dissociated nature).
4. No motor or sensory disability in the lower limbs.

CLINICAL PICTURE OF EPICONUS LESION: (L4,5 S1,2 SEGMENTS) :

1. Weakness or paralysis in the lower limbs, in the muscles supplied by $L_{4,5}$ and $S_{1,2}$ (dorsiflexors and plantar–flexors of the ankle and toes, the flexors of the knee and the extensors of the hip).
2. The ankle reflex is absent while the knee reflex is intact.
3. Sensory loss from L_4 to S_2 segment (usually of a dissociated nature).
4. Bladder disturbances may occur in the form of precipitancy.

N.B.: L.M.N. weakness of the ankle muscles (L4,5, S1,2) may be due to:

1. Radicular (cauda equina) or segmental (epiconus) lesion; in this case the gluteus maximum (L5, S1,2), tested by hip extension, is weak.
2. Peripheral nerve lesion; in this case the gluteus maximus is intact.



NEUROGENIC BLADDER

INNERVATION OF THE HUMAN URINARY BLADDER:

The urinary bladder, like any other viscus in the body is innervated through the autonomic system, and it has both a parasympathetic and a sympathetic supply.

1) **Parasympathetic supply**: it is from the 2nd, 3rd and 4th sacral segments. Its function is to contract the bladder wall and relax the sphincter.
2) **Sympathetic supply**: it is from 10th, 11th and 12th thoracic and 1st and 2nd lumbar segments. Its function, in animals, is to relax the bladder wall and to contract the sphincter, but in the human being, the sympathetic supply has no active role in the act of micturition.

Thus, the main nerve supply concerned with the act of micturition in the human being is the parasympathetic.

CONTROL OF THE ACT OF MICTURITION:

The wall of the urinary bladder contains certain receptors (stretch and chemical) which are stimulated by fullness of the bladder and acidity of the urine. Efferent impulses are carried by $S_{2,3,4}$ sensory parasympathetic fibres to the 2nd, 3rd and 4th sacral segments of the spinal cord. Efferent impulses carried by $S_{2,3,4}$ motor parasympathetic fibres lead to contraction of the bladder wall, relaxation of the sphincter, and its evacuation (in infants).

The afferent impulses reaching the sacral part of the cord ascend in the posterior column to reach the brain where the sensation of fullness of the bladder is perceived. Efferent impulses descend from a special bladder cortical centre for the conscious control of micturition. This centre is placed on the medial surface of the cerebral hemisphere in front of the foot area. This centre is developed with myelination of the pyramidal tract after the age of one year. The efferent impulses descend from this centre to the 2nd, 3rd and 4th sacral segments concerned with micturition through the pyramidal tracts. These efferent impulses are inhibitory in nature. Micturition takes place, in normal human beings, only when this pyramidal inhibition is released. This supraspinal inhibition is absent in infants below one year because of the absence of myelination of the pyramidal tracts, and the immaturity of the cortical bladder centre.

LESIONS AFFECTING BLADDER FUNCTION:

I. Lesions at the Level of the Reflex Arc (L.M.N.L.):

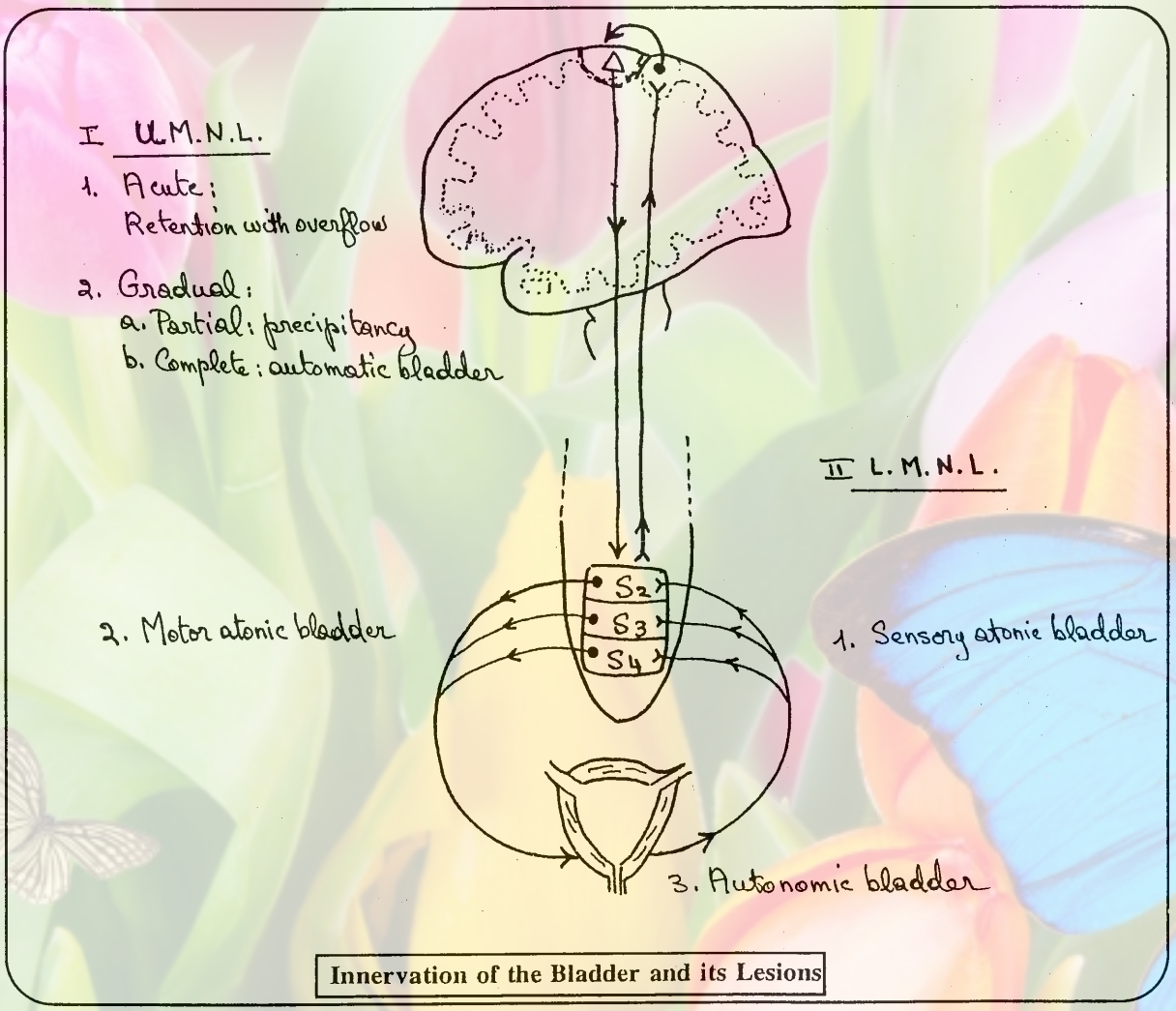
1. Lesions in the afferent fibres – **Sensory Atonic Bladder** characterised by:
 - Absence of the sense of fullness of the bladder.
 - Retention of urine associated with a huge size of the bladder.
 - Dribbling of urine every now and then because of overflow.

2. Lesion in the efferent fibres – **Motor Atonic Bladder** characterised by:
 - Preservation of the sense of fullness of the bladder.
 - Retention of urine associated with a moderate size of the bladder.
 - Inability to evacuate the bladder voluntarily.
 - Catheterisation is usually quickly done.
3. Lesion in both afferent and efferent fibres or in the spinal center – **Autonomic** or **Autonomous bladder** characterised by:
 - **Incomplete Irregular, Involuntary**, evacuation of the bladder. As the evacuation of the bladder depends on its myogenic contraction.

II. Lesions Above the Level of the Reflex Arc (U.M.N.L.):

1. Acute: **Retention with overflow**.
2. Gradual:
 a) Partial lesion: **Precipitancy** of micturition.
 b) Complete lesion: **Automatic Bladder**: this is characterised by complete, regular evacuation of the bladder which works by the spinal reflex arc.

N.B.: For any disturbance in bladder function (due to a nervous lesion) to occur, the lesion should be bilateral.



Innervation of the Bladder and its Lesions

PERIPHERAL NEURITIS

DEFINITION:

It is inflammation or degeneration of the peripheral nerves and/or the cranial nerves resulting in impairment of the conductivity of these nerves leading to motor, sensory and autonomic manifestations.

It is better to use the term "neuropathy" than "neuritis" as the pathological lesion may be degenerative and not always inflammatory.

CLASSIFICATION OF NEUROPATHY:

1. Mononeuropathy: affecting a single nerve trunk in one limb.
2. Mononeuropathy multiplex: affecting more than one nerve trunk in one limb.
3. Polyneuropathy: systemic affection of the peripheral nerves of all limbs.

CAUSES OF MONONEUROPATHY:

1. Trauma: wrong injection into a nerve, callus compression.
2. Infective: leprosy, herpes zoster.
3. Vascular: poly arteritis nodosa.
4. Metabolic: diabetes mellitus.

CAUSES OF POLYNEUROPATHY:

I) Heridofamilial:

1. Hypertrophic interstitial polyneuropathy.
2. Peroneal muscle atrophy.
3. Refsum's disease.

II) Symptomatic:

1. **Infective:**
 a) Viral: acute post–infective polyneuritis, mumps, measles.
 b) Bacterial: diphtheria, typhus, typhoid, tetanus.
 c) Mycobacterial: leprosy.
2. **Toxic:**
 a) Inorganic: all heavy metals (lead, arsenic, . . .).
 b) Organic: alcohol, insecticides.
3. **Nutritional**: beri beri, pellagra, S.C.D.
4. **Metabolic**. diabetes mellitus, uraemia, porphyria, amyloidosis.
5. **Endocrinal**: acromegaly, myxoedema.
6. **Auto–immune:**
 - Collagen diseases as rheumatoid arthritis, polyarteritis nodosa, scleroderma and systemic lupus erythematosis.
 - Landry–Guillain–Barre syndrome (acute post–infective polyneuropathy).
7. **Iatrogenic**: (P.N. 2ry to the use of drugs):
 I.N.H., cycloserine, sulphonamides, corticoids, phenytoin, vincristine.
8. **Neoplastic**: bronchial carcinoma, lymphoma, myeloma.

CLINICAL PICTURE OF POLYNEUROPATHY:

It will present by the 3 following manifestations in difference combinations.

A. Motor:

- Weakness or paralysis of L.M.N. nature (wasting, hypotonia, hyporeflexia . . .).
- The weakness and wasting are:
 - Bilateral and symmetrical.
 - Affecting L.L. more than U.L.
 - Affecting distal muscles more than proximal muscles.
 - Affecting extensors more than flexors.
- The weakness in the extensors of the distal group of muscles leads to bilateral foot drop and wrist drop.
- The ankle reflex is lost while the knee reflex is preserved.
- The cranial nerves may be affected specially Cr. III, VI, VII and X.
- The gait is high steppage due to the foot drop.

B. Sensory:

- Subjectively there is pain and paraesthesia in the limbs, specially distally.
- Objectively there is:
 1. Superficial sensory impairment of the stock and glove nature.
 2. Deep sensory loss specially distally with absence of deep reflexes.



C. Autonomic:

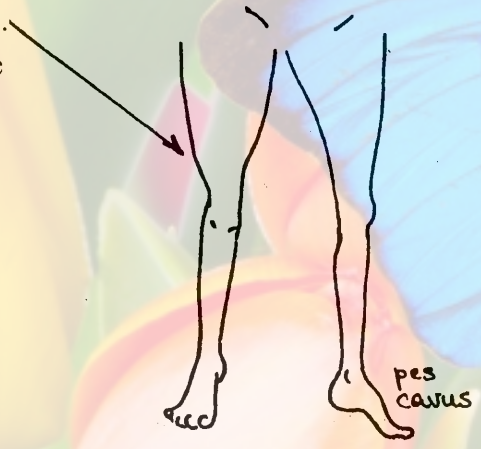
- Vasomotor: coldness and cyanosis of the limbs.
- Cutaneous: loss of hair, brittle nails, trophic ulcers.

N.B.: Regardless of the cause of the polyneuropathy, the clinical picture is essentially the same; variations depend on whether the motor, sensory or autonomic features predominate.

SPECIFIC TYPES OF POLYNEUROPATHY

PERONEAL MUSCLE ATROPHY

- A herido-familial type of P.N. appearing during the 1st and 2nd, decades of life.
- It has a gradual onset and a very slow, progressive course.
- The wasting and weakness start in the lower limbs in the peronii muscles then the anterior tibial group, then ascend to involve the muscles of the lower 1/3 of the thigh resulting in the **inverted champagne-bottle appearance.**
- In spite of the marked degree of wasting there is mild disturbance of motor power (i.e. discrepancy between the degree of wasting and the degree of motor weakness).
- Sensations are impaired specially the vibration sense which is markedly diminished.
- Skeletal deformities are usually present e.g., pes cavus.
- It may be associated with herido-familial ataxia.



REFSUM'S DISEASE

- This is a herido–familial lipid storage disease.
- The onset is during the 1st and 2nd decades.
- There is **hypertrophic neuropathy** associated with night–blindness, cerebellar ataxia, retinitis pigmentosa, ichthyosis (scaly skin) nerve deafness and skeletal deformities.
- There is accumulation in the liver and other tissues, of **phytanic acid**, due to lack of the enzyme phytanic acid hydroxylase which is necessary to metabolise phytanic acid, as a result serum phytanic acid is markedly elevated.
- Treatment is by plasmapheresis and by restriction of phytanic acid in the diet.

DIABETIC NEUROPATHY

Pathogenesis:

1. Diabetic microangiopathy of the vasa nervosa.
2. Ischaemia of the nerves, 2ry to atherosclerosis of the vasa nervosa.
3. Nutritional 2ry to hypovitaminosis (vit B_1, B_6 and B_{12}) due to the polyurea.
4. Metabolic due to the production of toxic ketonic bodies, leading to nerve damage.

Clinical picture:

- In early diabetes or in the pre–diabetic stage, the neuropathy is of the mononeuritic type which may affect the sciatic, femoral, lateral popliteal, ulnar or median nerves or Cr. III, VI or VII nerves.
- In frank diabetes, the neuropathy is of the polyneuritic type.
- The polyneuropathy is mainly **sensory** with pain and paraesthesia specially in the L.L., followed by superficial sensory loss of the stock and glove type. The deep sensations are also lost early in the disease, resulting in loss of deep reflexes and sensory ataxia.
- The muscle sense is increased at first resulting in **tender calf** followed later on by lost muscle sense.
- Motor weakness may occur late in the disease.
- Autonomic manifestations:
 - Impotence.
 - Sensory, motor or autonomic bladder.
 - Postural hypotension.
 - Silent myocardial infarction.
 - Gastroparesis diabeticorum: indigestion and delayed gastric emptying.
 - Hyperhydrosis or anhydrosis.
 - Trophic skin changes: ulcers, loss of hair, brittle nails, Charcot's neuropathic joint.

Treatment:

1. Proper management of diabetes: diet, oral hypoglycaemic drugs or insulin.
2. Vasodilators as piribedil (Trivastal)50 mg daily .
3. Capillary modulators: Ca Dobesilate (Doxium).
4. Vitamins B_1 100 mg daily, B_6 200 mg daily, B_{12} 1000 µg daily and A.T.P.(Adenoplex).
5. Carbamazepine (Tegretol) 200 mg twice daily or Gabapentine (Neurontine) 400 mg t.d.s. for the neuropathic pains.
6. Physiotherapy if motor weakness is present.

ACUTE INFECTIVE POLYNEURITIS
Landry–Guillain–Barré Syndrome

Aetiology:

It is due to an allergic or auto–immune reaction secondary to a previous non–specific virus infection.

Clinical Picture:

1. **Febrile stage**: it starts with an influenza–like attack with fever, headache, malaise, pains all over the body with no nervous symptoms.
2. **Latent stage**: the above symptoms disappear and the patient is free for 1–4 weeks.
3. **Paralytic stage**:
 - There is acute severe weakness or paralysis starting in the L.L. and ascending to involve the trunk and respiratory muscles, followed by the U.L. muscles.
 - Contrary to other types of polyneuropathy, the weakness is **proximal** more than distal.
 - In spite of the severe degree of paralysis, wasting is not present early in the disease.
 - Sensory impairment may occur resulting in stock and glove hyposthesia and deep sensory loss.
 - Early in the disease there is tenderness of the calves.
 - The cranial nerves are usually involved specially Cr. VII and X resulting in bilateral facial paralysis and bulbar symptoms.
 - **C.S.F. shows cytoalbuminous dissociation**.
 - Prognosis is good in 85% of the cases where recovery occurs in 3–4 weeks.

Treatment:

1. Absolute rest in bed till the heart rate reaches below 100/min. (i.e., return of the vagal tone).
2. Care of the bulbar muscles by:
 - Frequent suction of secretions from the pharynx.
 - Neostigmine l mg hourly I.M.
 - Tube feeding in case of pharyngeal paralysis.
3. Care of the respiratory muscles by:
 - Frequent suction to keep a patent airway.
 - Tracheostomy may be needed.
4. Corticosteroids (Prednisone or Prednisolone 60 mg/day).
5. Gamma–globulins and plasmapheresis: the most recent and effective treatment.
6. Vitamins B_1, B_6, B_{12} (Trivarol, Tri–B), I.M. daily.
7. A.T.P. 25 mg daily I.M. (Adenoplex).
8. Antibiotics e.g., Tetracycline to guard against 2ry infection.
9. Physiotherapy: Massage, passive and active exercises, proper positioning and electrical stimulation.

DIPHTHERITIC NEUROPATHY

Aetiology: Neuritis is the commonest and most important nervous complication of diphtheria. The exotoxin of Corynebacterium diphtheria has an affinity for the nerves.

Clinical Picture:

- Symptoms and signs occur 2–8 weeks after the appearance of diphtheria.
- Two main types of neuropathy may occur:
 1. Localised type: affecting Cr. III, VII and X nerves. The vagal paralysis results in bulbar symptoms with lost palatal and pharyngeal reflexes.
 2. Generalised type: affecting the peripheral nerves. The neuropathy is mainly motor.

Treatment: Antidiphteritic serum 100,000 Units I.M.

LEPROTIC NEUROPATHY

Aetiology:

- Causative organism: Mycobacterium leprae.
- The nerves are infiltrated by a granuloma (leprous nodule), leading to their thickening, degeneration and subsequent impairment of conductivity.

Clinical Picture:

- The neuropathy may be of the mono–or polyneuritic types.
- The commonest nerves affected are the lateral popliteal, ulnar, greater auricular, trigeminal and facial nerves.
- There is irregular thickening of the affected nerves.
- The neuropathy is mainly sensory.
- There are 3 clinical types:
 1. **Nodular (lepromatous)** leprosy: cutaneous nodules over the face and neck which ulcerate and heal by fibrosis leading to the **leonine facies**, usually associated with fever and lymphoadenopathy.
 2. **Maculo–anaesthetic (tuberculoid)** leprosy: maculo–anaesthetic skin patches with frequent trophic changes in the limbs (ulceration, gangrene).
 3. **Mixed** type.

Treatment:

- Sulphones: Dapsone is the best 100 mg/week orally.
- Diphenylthiourea: (Ciba 1906) l gm/day orally.
- Rifampicin: 500 mg/day, orally.

DEFICIENCY DISEASES

1. PELLAGRA

AETIOLOGY:

This is a multiple deficiency disease due to vitamin B complex deficiency specially **niacin**; this may result from:

1. Deficient intake of vit B or of proteins of high biological value (containing nicotinic acid or its precursor tryptophan).
2. Deficient absorption as in gastro–enteritis.
3. Parasitic infestation.

Clinical Picture

I. Cutaneous manifestations:

- They start as a bilateral and symmetrical dermatitis followed by hyperkeratosis (rough, scaly and desquamated skin) and pigmentation.
- The lesions involve the exposed areas (face, neck, dorsum of the hands and feet) and over bony prominences (trochanters, elbows, knees, heels).

II. Gastro–intestinal manifestations:

- Mouth: stomatitis, glossitis with atrophy of the papillae (glazed tongue).
- Stomach: dyspepsia, nausea, vomiting, epigastric pain.
- Intestines: colic, diarrhoea.
- Hepato–splenomegaly may be present.



III. Neuropsychiatric manifestations:

1) Peripheral neuropathy mainly sensory due to P.N. degeneration.
2) Paraplegia or quadriplegia (systemic paraplegia) due to pyramidal tract degeneration (pellagral lateral sclerosis).
3) Mentality changes as anterograde amnesia, depression, dementia and suicidal tendencies due to cerebral cortex degeneration.



Treatment:

1. Treatment of the cause e.g. parasitic infestation or gastro–enteritis.
2. Good diet, rich in animal proteins and vitamins.
3. Vit. B complex (the cheapest source is yeast) iron supplement (to correct anaemia).
4. Nicotinamide 500 mg I.V. daily in severe cases, followed after improvement by 200 mg daily I.V. for 2 weeks; maintenance dose 100 mg t.d.s. orally.

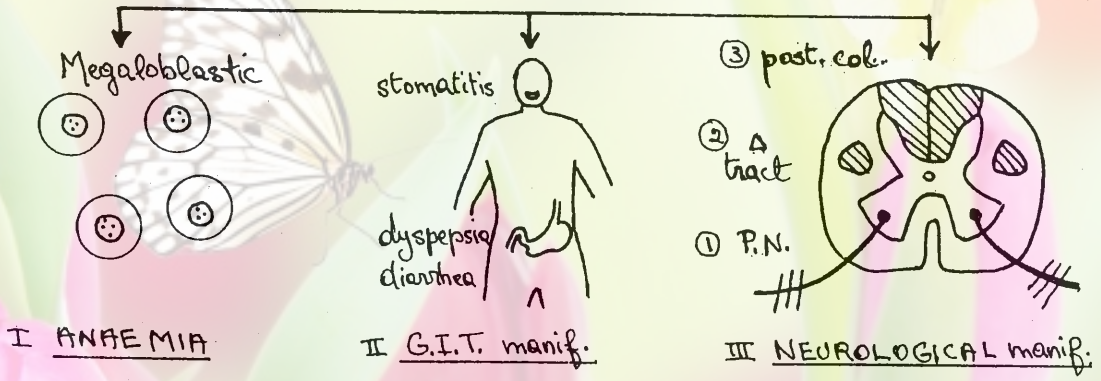
II. SUBACUTE COMBINED DEGENERATION (S.C.D.)

Aetiology:

This syndrome is due to **vit B_{12}** (cyanocobalamin) deficiency. This may result from:

1. Deficient intake (rare).
2. Deficiency of the intrinsic factor essential for vit B_{12} absorption:
 * Pernicious anaemia. * Atrophic gastritis, cancer stomach, gastrectomy.
3. Deficient absorption as in malabsorption syndrome or parasitic infestation.
4. Increased demands in infancy and pregnancy.
5. Hepatic failure.

Clinical Picture: It presents with a triad of:



I. Anaemia:

* There is pallor, fatigue, palpitation, dyspnoea.
* It is of the megaloblastic hyperchromic type, with anisocytosis and poikilocytosis.

II. Gastrointestinal manifestations:

* Mouth: stomatitis with a red smooth and swollen tongue (beefy tongue).
* Stomach: dyspepsia, nausea, vomiting, achlorhydria.
* Intestines: colic, diarrhoea.

III. Neurological manifestations:

There is combined degeneration of the peripheral nerves, pyramidal tracts and posterior columns.

1) Pains and paraesthesias in the limbs followed by stock and glove hyposthesia. The ankle reflex is lost while the knee reflex is preserved due to P.N. degeneration.
2) The pyramidal degeneration results in weakness in the limbs with +ve Babinski sign; however the tone and deep reflexes are diminished (due to the peripheral nerve and post, column lesions).
3) Deep sensory loss (sensory ataxia) due to posterior column degeneration.

TREATMENT:

1. Vit B_{12} 1000 µgm I.M. daily for 2 weeks then 100 µgm twice per week.
2. Liver extract. 15 units twice per week and vit. B complex.
3. Gastric enzymes and dilute HCL. 4–8 cc with meals.

III. BERI–BERI

Aetiology: A disease 2ry to thiamine (**vit. B_1**) deficiency affecting the nervous system (dry beri–beri) or the cardiovascular system (wet beri–beri).

Clinical Picture:

1. Dry beri–beri: peripheral sensory neuropathy.
2. Wet beri–beri: picture of congestive heart failure.
3. Cerebral type (Wernicke's encephalopathy): amnesia, ophthalmoplegia, nystagmus & ataxia.

TREATMENT:

1. Thiamine 100 mg daily and vit. B complex.
2. Diet rich in vitamins and low in salt.
3. Digitalisation and diuretics in case of heart failure.

N.B.:

CAUSES OF MOTOR NEUROPATHY :

1. Lead neuropathy.
2. Diphtheritic neuropathy.
3. Acute infective polyneuritis.
4. Porphyria.

CAUSES OF SENSORY NEUROPATHY :

1. Diabetic neuropathy.
2. Alcoholic neuropathy.
3. Vitamin deficiency neuropathy.
4. Arsenic neuropathy.
5. Leprotic neuropathy.

CAUSES OF TENDER CALF MUSCLES :

1. Deep venous thrombosis (D.V.T.).
2. Landry–Guillain–Barre syndrome.
3. Myositis.
4. Early diabetic neuropathy.

CAUSES OF THICKENED NERVES :

1. Interstitial hypertrophic P.N.
2. Refsum's neuropathy.
3. Leprotic neuropathy.
4. Neurofibroma.
5. Acromegaly.
6. Myxoedema.
7. Amyloidosis.

CAUSES OF LOST ANKLE REFLEX WITH PRESERVED KNEE REFLEX

1. Epiconus lesion.
2. Cauda equina lesion affecting S1 root.
3. Peripheral neuropathy.
4. Subacute combined degeneration.
5. Friedreich's ataxia.

MYOPATHIES

DEFINITION: Myopathies are a group of diseases of the skeletal muscles characterised by gradual progressive degeneration of these muscles. When this degenerative process is genetically determined, it is termed "progressive muscular dystrophy."

CLINICAL PICTURE:

It usually affects the 1st and 2nd decades; the onset is gradual and the course progressive.

SYMPTOMS:

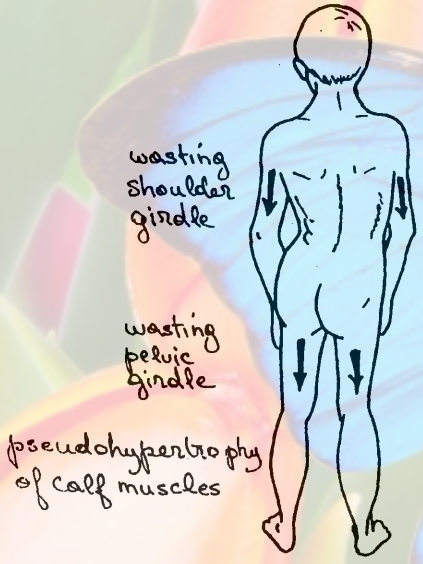
1. Clumsy gait.
2. Inability to climb the stairs.
3. Protuberant abdomen.
4. Weakness and wasting of certain muscles (shoulder and pelvic girdles and trunk).

SIGNS:

1. Weakness of the skeletal muscles of the body especially those which develop early during intra–uterine life i.e., the trunk, shoulder and pelvic girdle muscles.
2. The weakness is of L.M.N. nature i.e. it is associated with wasting, hypotonia, hyporeflexia . . .
3. The weakness and wasting are bilateral, symmetrical and proximal more than distal i.e., the shoulder and arm are more affected than the forearm and hand and the hip and thigh are more affected than the leg and foot.
4. The weakness and wasting of the shoulder, pelvic girdle and trunk muscles results in:
 a) **Winging of the scapulae** due to weakness of the serratus anterior and trapezius.
 b) **Pot–belly abdomen** due to weakness of the abdominal muscles.
 c) **Exaggerated lumbar lordosis** due to weakness of the extensor muscles of the trunk in an attempt from the patient, to prevent himself from falling forwards by the effect of gravity.
 d) **Waddling gait** due to weakness of the gluteus medius & minimus (abductors of the hip).
 e) Characteristic manner in getting up from the floor (climbing test or **Gower's sign**) due to weakness of the gluteus maximus.



GOWER'S SIGN



5. There is **selectivity** of the involved muscles e.g., there is atrophy of the sternal head of the pectoralis major with preservation of its clavicular head; the sternomastoid muscle is also spared.
6. The facial muscles are bilaterally weak in certain cases (facio–scapulo–humeral type).
7. **Pseudo hypertrophy** of the muscles is present in certain cases (Duchenne type) affecting mainly the gluteus maximus, quadriceps and calf muscles in the L.L., and the deltoid, supra and infra spinatus muscles in the U.L.
8. Later on there are fibrosis and contractures of the affected muscles resulting in **skeletal deformities** e.g. pes cavus and talipes equinus.
9. E.C.G. changes, histological changes in the heart and cardiomyopathy may be found specially in Duchenne type.
10. No sensory changes, fasciculations or sphincteric disturbances.

CLINICAL TYPES OF PROGRESSIVE MUSCULAR DYSTROPHIES:

1. **Shoulder girdle types**:
 a) Scapulo–humeral type (Erb).
 b) Facio–scapulo–humeral type (Landouzy and Dejerine).
2. **Pelvic girdle types**:
 a) Pseudo–hypertrophic type: (severe Duchenne subtype starting in the first decade of life, and the benign Becker subtype starting in the 2nd and 3rd decades of life).
 b) Atrophic type (Leyden–Mobius).
3 **Other rare types**:
 a) Distal type of Gower. b) Ocular type. c) Oculo–pharyngeal type.

	Duchenne	Becker
1. Age of onset	1st decade	2nd and 3rd decades
2. Course	Progressive	Slowly progressive
3. Skeletal deformities	Present	Absent
4. E.C.G. changes	Commonly present	Absent

GENETICALLY-DETERMINED MUSCULAR DYSTROPHIES can be classified into:

1. X–linked pseudohypertrophic types: There is a major mutation of a large gene located on the short arm of the X chromosome, leading to deficiency of a protein "dystrophin" resulting in:
 a) Severe form "Duchenne subtype." b) Benign form "Becker subtype"
2. Autosomal dominant: Facio–scapulo–humeral type.
3. Autosomal recessive: Limb girdle type "Erb's and Leyden–Mobius."

OTHER CAUSES OF MYOPATHY:

1. Endocrinal: Thyrotoxicosis, acromegaly, Cushing's syndrome.
2. Carcinomatous: Bronchogenic carcinoma, ovarian tumours.
3. Metabolic: Periodic paralysis syndromes due to disturbance of K balance.
4. Iatrogenic: Corticosteroids, chloroquine, vincristine.
5. Toxic: Alcohol.

INVESTIGATIONS:

1. **Estimation of creatine and creatinine in urine:** Normally creatine is absent from urine and creatinine is present (about 22–23 mg/kg body weight/24 hours). However creatine is normally present in urine in children below 2 years and in pregnant females. In cases of myopathy creatine will appear in the urine and creatinine will decrease, because the diseased muscles cannot use their creatine and metabolise it to creatinine.
2. **Estimation of serum enzyme:** Serum aldolase, transaminases and creatine phosphokinase are highly elevated in the serum due to degeneration of the muscle fibres.
3. **Creatine tolerance test:** Administration of 2 g creatine by mouth is followed by excretion of excess amount of creatine in urine denoting lack of the ability to retain creatine and lack of the ability to convert creatine to creatinine in cases of myopathy.
4. **E.M.G.:** Diminished amplitude and duration of motor units, & early interference pattern.
5. **Muscle biopsy:** Degeneration of muscle fibres and its replacement by fibrous and fatty tissue.

TREATMENT: There is no specific treatment; management is supportive:

1. A.T.P. 25 mg daily I.M.
2. Vitamin E as it helps the retention of creatine in the body.
3. Physiotherapy in the form of massage, passive and active exercises.
4. Orthopaedic procedures e.g. tenotomy to correct contractures.
5. If the patient is bed–ridden, guard against pneumonia, bed–sores and D.V.T.
6. Genetic counselling: detection of the carrier state is essential for prophylaxis.

Causes of death in muscle dystrophies:

1) Paralysis of respiratory muscles.
2) Infections specially hypostatic pneumonia.
3) Cardiomyopathy in Duchenne type.

POLYMYOSITIS

DEFINITION: It is an acquired autoimmune disease of the skeletal muscle which may be precipitated by:

- Systemic infection.
- Collagen disorder e.g. SLE.
- Immunisation.
- Neoplasm.

CLINICAL PICTURE:

1. Age: usually above 30 years, unless there is an associated collagen disorder.
2. Onset: acute or subacute with general symptoms of fatigue followed by:
3. Pain and tenderness of the muscles (60% of cases).
4. Weakness affecting:
 - The proximal muscles of U.L. and L.L.
 - The posterior neck muscles (forward lolling of the head).
 - The pharyngeal and laryngeal muscles (bulbar symptoms).
5. No involvement of the ocular muscles.
6. The deep reflexes are intact.





N.B.: Dermatomyositis: In this disease the clinical picture of polymyositis is associated with cutaneous manifestations as violet discolouration of the areas of skin exposed to light, tightening of skin and telangiectasia.

INVESTIGATIONS:

1. Raised E.S.R.
2. Raised C.P.K. in serum.
3. E.M.G. shows a myopathic pattern with fibrillation.
4. Circulating antibodies (antinuclear factor, rheumatoid factor) may be found.

TREATMENT:

1. Prednisolone 60 mg daily in divided doses; once there is improvement, gradually reduce to a maintenance dose; if it is stopped too early relapse may occur.
2. Immunosuppressive drugs and Methotrexate or Azothioprine in resistant cases.
3. Treatment of cause, if present.

FAMILIAL PERIODIC PARAL YSIS SYNDROMES

These are acute recurrent attacks of weakness resulting from alterations in the serum potassium (K^+) levels of familial origin.

	Hypokalaemic periodic paralysis	Hyperkalaemic periodic paralysis
Age	2nd decade.	1st decade.
Ppting factor	– High carbohydrate meal. – Rest after exercise.	– Cold. – Exercise.
Onset	Usually on awakening in the morning.	Immediately after exercise or cold.
C.P.	Weakness starts in L.L. and rapidly becomes generalised. No bulbar/respiratory affection.	Weakness starts in L.L. & rapidly becomes generalised. Myotonia in some cases.
Duration	Hours to days.	Less than one hour.
Serum K^+	<3 mEq/L.	>4.5 mEq/L.
Treatment: Acute case Prophylactic	KCl I.V. drip (with ECG monitor) – Acetazolamide 250 mg/daily. – Low CHO, high K^+ diet.	Ca gluconate I.V. drip or glucose & insulin drip. – Thiazide diuretics.

MYASTHENIAS

I. PRIMARY MYASTHENIA: (Myasthenia gravis)

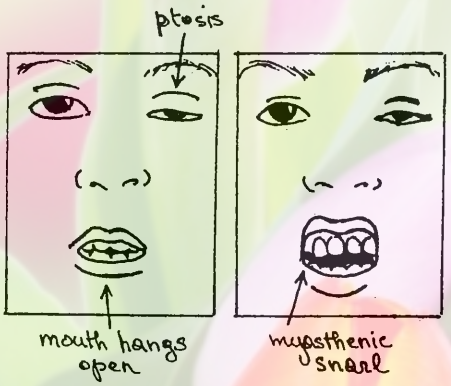
DEFINITION: It is a disorder of transmission at the neuromuscular junction, manifesting itself clinically by easy fatiguability of the skeletal muscles, specially on repetition of movement, which is relieved by rest.

AETIOLOGY:

- It is an auto–immune disease with production of antibodies against the acetylcholine receptors of the neuromuscular junction, leading to their destruction. These Acetylcholine Receptor antibodies (AChR antibodies) are found in the patient's serum.
- Normally the thymus gland produces T–cell lymphocytes which participate in immune responses. Thus, thymic dysfunction (80% of patients) may lead to disturbed immune responses with production of AChR antibodies.

CLINICAL PICTURE:

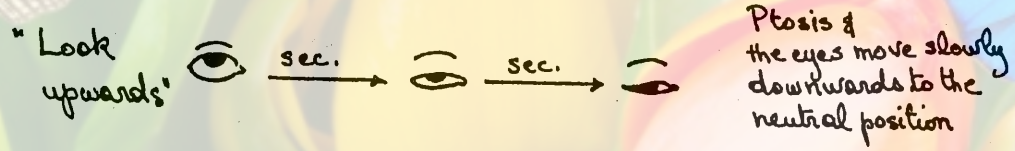
1. Age: usually 20–40 years. 2. Sex: more in females.
3. Onset and Course: usually gradual onset and progressive course.
4. It affects only the **skeletal muscles** of the body.
5. The disease is characterised by **easy fatiguability** of the muscles on repetition of movement.
6. The disease has a characteristic **descending march course** affecting the following muscles:
 - The **ocular muscles** leading to ptosis, diplopia & ophthalmoplegia. In some cases they may be the only muscles affected throughout the illness.
 - The **jaw muscles** leading to the mouth hanging open.
 - The **facial muscles** leading to an expressionless appearance. On smiling there is a characteristic smile (myasthenic snarl).
 - The **bulbar muscles** leading to the tetrad of bulbar symptoms (dysphagia, dysarthria, dysphonia and nasal regurge).
 - The **skeletal muscles:** the muscles of the U.L. are affected before those of the L.L. & the proximal muscles are affected more than the distal muscles.
 Weakness of the neck muscles leads to lolling of the head.



7. **Diurnal variation** is noted. The motor power is good in the early morning and is at its worst at the end of the day.

INVESTIGATIONS:

1. **Clinical tests:**
 a) Induction of fatigue by repetition of movement.



b) **Walker's test**: apply the sphygmomanometer cuff to the arm and raise the pressure above the systolic and ask the patient to do rapid repetitive movements with his hand till exhaustion occurs. Then release the cuff; if the case is myasthenia, ptosis with occur within 10 seconds.

2. **Pharmacological tests**:
 a) Prostigmine test: 2.5 mg prostigmine plus one ampoule atropine are injected I.M.; in myasthenia, improvement of the fatigued muscle will occur within $^1/_2$ hour.
 b) Tensilon (edrophonium) test: Tensilon is a short acting cholinergic drug, 2 mg are injected I.V. to observe for idiosyncrasy; if not, inject 8 mg. After one minute the ptosis will disappear and the fatigue will disappear if the case is myasthenia.
3. **Serological tests**: Acetylcholine receptor antibodies are detected in the serum of 90% of patients.
4. **E.M.G.**: Reduction in the amplitude of the compound muscle action potential evoked by repetitive supramaximal nerve stimulation (the decrementing response).



5. **Radiological tests**: Plain x–ray and C.T. tomography of anterior mediastinum and isotope (Gallium) scan in cases of suspected thymoma.
6. **Muscle biopsy**: increased lymphocytes (lymphorrhages).

TREATMENT:

1) Medical:

1. **Anticholinesterase drugs**: They interfere with cholinesterase, the enzyme responsible for the breakdown of acetylcholine, allowing increased receptor stimulation.
 a) Neostimgine (prostigmine): tablet 15 mg or ampoule $^1/_2$ mg I.M. Its action starts within $^1/_2$ hour and reaches its peak within 2 hours to fade away in 4 hours.
 Dose: start by one tablet t.d.s. or one ampoule t.d.s. & gradually increase the dose till improvement occurs.
 b) Pyridostigmine (Mestinon): tablet 60 mg, its action starts within 2 hours, reaches its peak in 4 hours to fade away in 8 hours.
 Dose: start by one tablet t.d.s. and gradually increase till improvement occurs.
 Thus we combine Prostigmine and Mestinon and give them to the patient every 8 hours.
2. **Steroids**: Prednisolone is used in patients not responding to anticholine esterase drugs; it may produce a remission which may last up to 5 years.
3. **Immunosuppressants** other than steroids e.g. Azathioprine or cyclophosphamide in patients not responding to steroids.

2) Surgical:

Thymectomy is indicated for young patients with generalised symptoms of less than 5 years duration.

3) Plasmapheresis:

The patient's plasma is "exchanged" for albumin. This reduces the acetylcholine receptor antibody levels. As the antibody level increases after 2–3 weeks several exchanges are required. Thus plasmapheresis is best indicated in:

1. Myasthenic crisis. 2. To improve the clinical state before thymectomy.

II. SECONDARY MYASTHENIA:

There is easy fatiguability of the skeletal muscles 2ry to a known cause e.g. bronchogenic carcinoma, polymyositis, lupus erythematosus . . . Contrary to 1ry myasthenia it does not have a march course, does not respond to prostigmine but responds to guanidine HCl which therefore is used for its treatment. Also contrary to 1ry myasthenia with successive stimulation of the muscle, during E.M.G. examination there is an increase of the amplitude of the motor unit potentials (Eaton–Lambert phenomenon); this is because repeated stimulation squeezes the vesicles at the neuro–muscular junction & releases the acetylcholine stored within them.

III. NEONATAL MYASTHENIA:

1. It occurs in the offspring of myasthenic women and is due to passive transplacental passage of (AChR antibodies).
2. There is poor crying and suckling and the infant is floppy.
3. The weakness may persist till the end of the 3rd month.
4. Treatment with anticholine esterases is required.

IV. CONGENITAL MYASTHENIA:

1. This is a very rare type which begins in infancy and persists throughout life.
2. It is due to structural abnormalities of the receptors at the N.M.J. Thus there are no AChR antibodies in the serum and thymectomy is contraindicated.

MYOTONIAS

DEFINITION: Myotonic phenomenon is delayed relaxation of the skeletal muscles after voluntary, mechanical or electrical stimulation.

N.B.:

- **Voluntary**: when the patient voluntarily clenches his fist, he is unable to open his hand, except after sometime.
- **Mechanical**: if we tap the thenar eminence, adduction of the thumb occurs with difficulty & delay in abduction. Similarly, if we tap the tongue a dimple is observed due to delayed relaxation of the muscles.
- **Electrical**: 2–3 milliamperes are sufficient to produce contraction of the muscle due to hyper-excitability (normally at least 6 milliamperes are required).



The Myotonic Phenomenon:

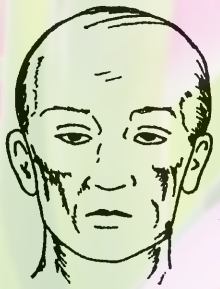
- **Improves by**: repetition of movement, warmth, calcium, quinine, procainamide.
- **Is worsened by**: cold, potassium and prostigmine.

TYPES OF MYOTONIA: Two main clinical types are known:

Features	Myotonia congenita	Myotonia artophica
1. Heredity	Herido–familial.	Usually sporadic.
2. Age	1st & 2nd decades.	3rd & 4th decades.
3. Sex	Females more than males.	Males more than females.
4. State of muscles	Pseudohypertrophy of all muscles of the body.	Atrophy specially of – Facial muscles. – Mastication muscles. – Sternomastoid. – Distal muscles of limb.
5. Dystrophic changes	Absent.	Present in the form of: cataract, frontal baldness & testicular atrophy.
6. E.C.G. changes	Uncommon.	Common.

Rare types of myotonia:

1. Myotonia acquisita, associated with myxoedema and hyperkalaemia.
2. Myotonia paradoxica, occurs with exercise.



TREATMENT:

1. Quinine: 5 grain t.d.s.
2. Procainamide: 500 mg t.d.s.
3. Calcium gluconate: 10–20 c.c. I.V. daily.
4. Potassium exchange resins.
5. Epanutin: 100 mg t.d.s.

N.B.: CAUSES OF PSEUDO-HYPERTROPHY OF MUSCLES:

1. Duchenne muscle dystrophy.
2. Becker muscle dystrophy.
3. Myotonia congenita.
4. Late cases of acromegaly.
5. Myxoedema.

N.B.: MOST COMMON CAUSES OF PES CAVUS:

1. Friedreich's ataxia
2. Duchenne myopathy.
3. Peroneal muscle atrophy.
5. Syringomyelia.

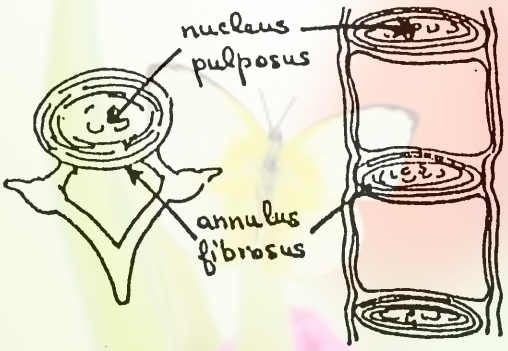
SPONDYLOSIS

DEFINITION:

It is the gradual, progressive degeneration of the intervertebral discs, specially those which are freely mobile as they are more subjected to the process of wear and tear. The freely mobile discs are mainly found in the cervical and lumbar regions.

PREDISPOSING FACTORS:

1) Old age.
2) Diabetes mellitus.
3) Excessive mobility of the spine as in labourers.



PATHOLOGY:

The intervertebral disc is formed of a central gelatinous part, "the nucleus pulposus," surrounded by a fibrous tissue ring, "the annulus fibrosus" and covered from above and below by a cartilaginous plate.

In spondylosis there is degeneration of the annulus fibrosus, leading to herniation of the nucleus pulposus with subsequent compression of adjacent structures. As the weakest parts of the annulus fibrosus are the lateral and posterior parts, the herniation will be either lateral, posterior or posterolateral.



1. Lateral Prolapse
2. Posterior Prolapse
3. Postero-lateral Prolapse

CLINICAL PICTURE:

1. CLINICAL PICTURE OF CERVICAL SPONDYLOSIS:

It may present by one of the 3 following manifestations depending on the direction of prolapse of the disc.

1) **Manifestations of root compression** (***lateral prolapse***):

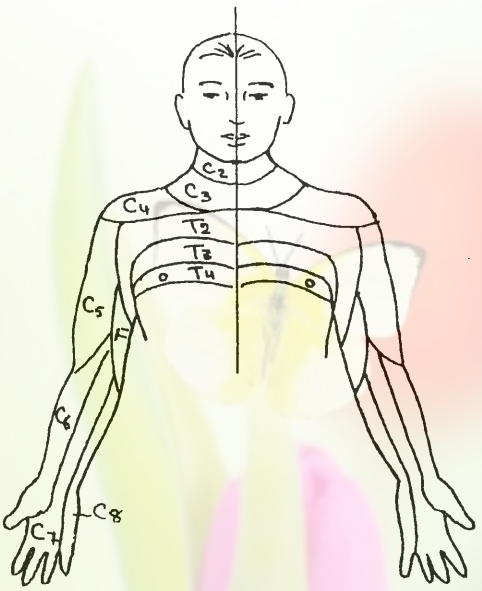
a) **Anterior root compression**: This results in a localised L.M.N. weakness or paralysis in the muscles supplied by the compressed root:

Root	Action	Muscle
$C_{1,2}$	Lateral movement of neck	Sternomastoid & trapezius
$C_{3,4}$	Elevation of shoulder	Supra and infraspinatus
C_5	Abduction of shoulder	Deltoid
$C_{5,6}$	Flexion of elbow	Biceps & brachioradialis
$C_{6,7}$	Extension of elbow	Triceps
$C_{7,8}$	Extension of wrist	Extensors of wrist
C_8,T_1	Flexion of wrist & movement of small ms of hands	Flexors of wrist

b) **Posterior root compression:**

- This results in pain and paraesthesias at the onset, due to posterior root irritation, which are referred to the shoulder and upper limbs (brachial neuralgia).
- Later on, there is hyposthesia or anaesthesia in the dermatome supplied by the compressed root:

Root	Sensory distribution
C_2	Lateral aspect of neck
$C_{3,4}$	Shoulder down to manubrium anteriorly
C_5	Lateral aspect of arm
C_6	Lateral aspect of forearm, thenar eminence & thumb
C_7	Middle aspect of forearm, middle of palm, middle 3 fingers
C_8	Medial aspect of forearm, hypothenar eminence & little finger
T_1	Medial aspect of arm



2) **Manifestations of cord compression** (***Postero–lateral prolapse***)**:**

a. Compression of the pyramidal tracts: This results in weakness or paralysis with signs of U.M.N.L. i.e. hypertonia, hyperreflexia. below the level of compression.

b. Compression of the spino–thalamic tracts: This results in superficial sensory loss below the level of compression.

c. Compression of the posterior column: This results in deep sensory loss i.e. sensory ataxia below the level of compression.

3) **Manifestations of root and cord compression:**

a. In the upper limb there will be combined signs of L.M.N. lesion in the form of wasting of muscles with signs of U.M.N.L. in the form of hypertonia and hyperreflexia, i.e. **tonic atrophy**.

b. In the lower limbs there will be weakness with signs of U.M.N. lesion.

N.B.: INVERTED SUPINATOR REFLEX:

In cases where a lesion involves the fifth cervical segment, a pathological reflex, known as the inverted supinator reflex can be elicited and is characterised by:

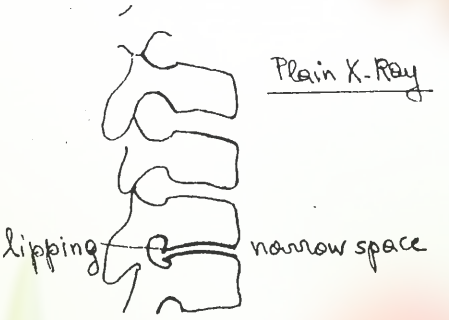
1) Lost or weak biceps reflex.
2) Exaggerated triceps reflex.
3) On eliciting the brachioradialis or biceps reflex, finger flexion will occur instead of flexion at the elbow; this is due to irradiation of nerve impulses in the spinal cord with stimulation of the A.H.C. of C_8 and Th_1 segments.

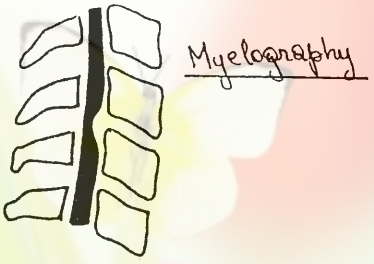
II. CLINICAL PICTURE OF LUMBAR SPONDYLOSIS:

This will present with the same picture of Cauda equina lesion.

INVESTIGATIONS OF SPONDYLOSIS:

1) Plain X–ray:
 1. Narrowing of the intervertebral disc spaces.
 2. Sclerosis of the adjacent surfaces of the vertebrae.
 3. Lipping or osteophytic formations due to calcification of the prolapsed parts and ligaments
 4. Straightening of the spines (loss of lordosis).
2) C.T. scan and M.R.I. show small disc protrusions missed by other methods of investigations
3) Myelography shows an anterior filling defect due to disc protrusions and compression of the cord.





TREATMENT OF SPONDYLOSIS:

I. Medical:

- Analgesics as Voltaren, and muscle relaxants as Idarelax.
- Anabolics, vasodilators and multi–vitamins may be used.

II. Physiotherapy:

- Short wave therapy to the neck.
- Plastic neck collar which if used, should never be applied for more than 3 months to avoid fibrosis of the muscles.
- Traction of the vertebrae often relieves the pain.

III. Surgical: Decompression by laminectomy. Surgical interference is indicated in cases of:

1) Failure of the medical treatment.
2) Severe intolerable pain.
3) Bladder disturbances (in cases of cervical spondylosis), denoting severe cord compression.

BRACHIAL NEURALGIA: It is severe radicular pain along the distribution of the brachial plexus (C5 → Th1) i.e. in the shoulder and U.L. It may be due to:

1. Cervical spondylosis and disc prolapse.
2. Cervical rib (p. 97).
3. Pancoast tumour.
4. Brachial neuritis: a mononeuritis multiplex of acute onset due to infection or post–vaccine. Severe pain is followed by weakness of U.L.; wasting occurs within 1 month. The pain is not related to neck movements.
5. Referred pain from the heart (angina, myocardial infarction), or gallbladder.

N.B.: The Carpal Tunnel Syndrome only causes pain & paraesthesias in the hand due to compression of the median nerve in the tunnel, the shoulder & arm are unaffected (p. 97).

SCIATICA

DEFINITION: It is radicular pain along the distribution of the sciatic nerve ($L_{4,5}$ $S_{1,2,3}$) i.e. along the back of the thigh, leg and foot.

CAUSES:

I. In the **spinal canal** at the lumbosacral regions:
 1) Acute lumbar disc prolapse.
 2) Lumbar spondylosis.
 3) Fracture or dislocation.
 4) Pott's disease or tumours.

II. In the **intervertebral foramina**:
 1) Neurofibroma.
 2) Radiculitis.
 3) Ankylosing spondylitis.

III. In the **pelvis**: compression of sciatic plexus over the sacro–iliac joint by:
 1) Malignant tumours of the bladder, rectum, ovaries . . .
 2) Pelvic abscess.
 3) Pregnant retroverted uterus.

IV. In the **sciatic nerve**:
 1) Neuritis as diabetic, alcoholic and rheumatic.
 2) Pressure on the nerve by dislocated head of femur.
 3) Wrong injection into the nerve.

V. **Referred** sciatic pain 2ry to hip or sacro–iliac joint disease.

The most common causes of sciatica are: • Acute disc prolapse • Lumbar spondylosis.

Acute disc prolapse: There is sudden rupture of the annulus fibrosis followed by bulging (herniation) of the nucleus pulposus; this compresses the spinal roots. It may occur at any age and usually follows trauma, as lifting heavy objects or jumping to the floor from a height. Plain x–ray of the back shows narrowing of the intervertebral spaces with no degenerative changes.

	Acute disc prolapse	Lumbar spondylosis
Age	Any age	Middle and old age
Cause	Traumatic	Degenerative
Onset	Acute	Gradual
X–ray	Narrowed intervertebral space	Narrowed intervertebral space Sclerosis, lipping & osteophytes

CLINICAL PICTURE OF SCIATICA:

I. Symptoms

Pain and paraesthesias along the course of the sciatic nerve, aggravated by walking which stretches the nerve and also by coughing, straining or sneezing. The pain is relieved on bed rest in cases of disc lesions but it is not relieved and may even worsen in cases of intraspinal space–occupying lesions.

II. Signs

1) Sensory: Tenderness and discomfort on direct pressure on the sciatic nerve.
2) Motor: Slight L.M.N. weakness in the muscles supplied by the nerve and the ankle reflex may be weak or lost.
3) Back signs: The paravertebral muscles are spastic; resulting in loss of lumbar lordosis, tenderness and limitation of movements of the spine; specially in cases of disc prolapse.
4) Signs of meningeal irritation: +ve Kernig, Lassegue & Brudzinski signs.

INVESTIGATIONS:

1) In cases with suspected lesion of the spinal canal:
 – Plain x–ray – Myelography – C.T. scan and M.R.I.
2) Rectal and vaginal examination 3) Plain x–ray for the hip joint.
4) Urine analysis and blood sugar curve.

TREATMENT:

1. Treatment of the cause.
2. In cases of disc prolapse.
 - Bed rest on a hard mattress for 2–3 weeks.
 - Analgesics (e.g. Voltaren) and muscle relaxants (e.g. Idarelax) to relieve the pain.
 - Traction on the affected leg often lessens the pain and speeds recovery.
 - A spinal brace may be used during the day to reduce mobility of the back and to relieve pain
 - The patient should never lift heavy objects or bend his back to pick objects from the floor but should bend his knees and keep his back straight
 - Decompression laminectomy when there is:

• Failure of medical treatment. • Frequent recurrence of pain.
• Development of a neurological deficit. • Inability to take a long bed rest.

MOTOR NEURONE DISEASE

Definition: It is a degenerative disease of a gradual onset and progressive course, affecting the motor system only (systemic disease). It may affect the U.M.N., or the L.M.N. or both.

Clinical Picture:

- Age of onset: usually the middle and old ages.
- Sex: males are more affected than females.
- Onset and Course: gradual onset and progressive course.
- Signs and symptoms: this depends on whether the U.M.N. or the L.M.N. or both are affected. As this is a systemic disease, the S & S are usually bilateral.

1. U.M.N. AFFECTION

a) In the spinal cord: Syndrome of "**Lateral Sclerosis**". This results in bilateral S & S of U.M.N.L., resulting in:
– Spastic paraplegia if the lesion is below the cervical region, or
– Spastic quadriplegia if the lesion is at the cervical region.
b) In the brain stem or the cerebral hemisphere: Syndrome of "**Pseudo–bulbar palsy**".
 1. Bulbar symptoms:
 – dysphagia. – nasal regurgitation.
 – dysarthria. – hoarseness of voice.

2. Quadriplegia & signs of U.M.N.L. in the upper & lower limbs.
3. Exaggerated palatal and pharyngeal reflexes.
4. Exaggerated jaw reflex (if the lesion is above the pons).
5. Emotional and mood changes may be present.

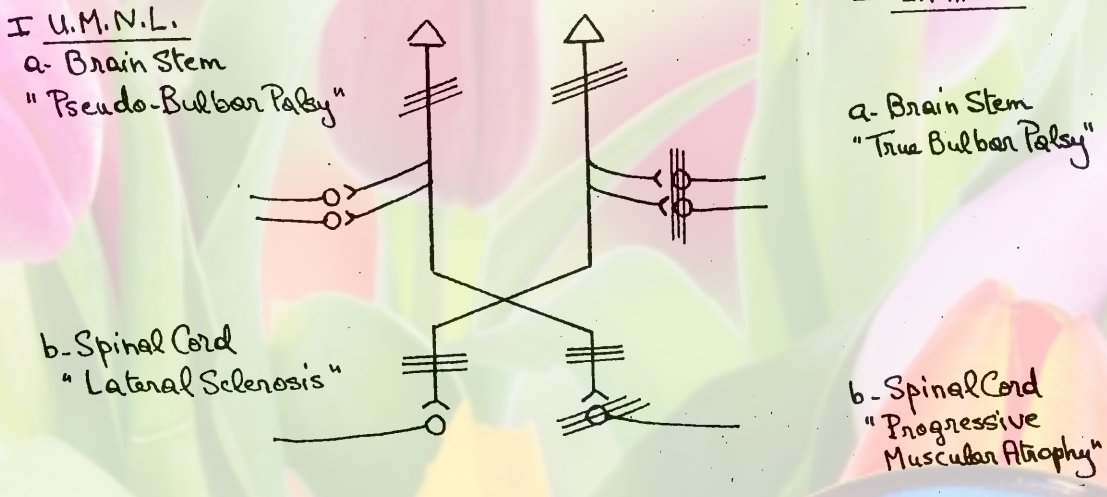
2. L.M.N. AFFECTION:

The disease has a tendency to affect the A.H.C. in the spinal cord or the cranial nerve nuclei in the brain stem.

a) In the A.H.C.: Syndrome of "**Progressive muscular atrophy**". The A.H.C, mostly affected are those of the lower cervical region and to a lesser extent those of the lumbar region, resulting in weakness with signs of L.M.N. lesion i.e. twsting, hypotonia, hyporeflexia and fasciculations. This clinical picture starts in the upper limbs, specially in the hands, then the forearms and shoulders. In the lower limbs the signs are late and of a much lesser extent.

b) In the cranial nerve nuclei: Syndrome of "**True bulbar palsy**" manifested by.
 1. Bulbar symptoms:
 – dysphagia. – nasal regurgitation.
 – dysarthria. – hoarseness of voice.
 2. Absent palatal and pharyngeal reflexes.
 3. The tongue is wasted and shows fasciculations.
 4. There is no quadriplegia, or emotional changes.

MOTOR NEURON DISEASE



Pseudo-bulbar palsy	True bulbar palsy
1. U.M.N. lesion.	• L.M.N. lesion.
2. Associated with quadriplegia.	• No quadriplegia.
3. Exaggerated palatal & pharyngeal reflexes.	• Lost palatal and pharyngeal reflexes.
4. The jaw reflex may be exaggerated.	• Absent jaw reflex.
5. Emotional lability may be present.	• No emotional lability.
6. Tongue: no wasting or fasciculations.	• Tongue: small, flaccid & shows fasciculations.

3. COMBINED U.M.N. AND L.M.N. AFFECTION:

Syndrome of **"amyotrophic lateral sclerosis":**

There are combined signs and symptoms of lower and upper motor neurone lesions:

1. In the upper limbs there will be weakness, associated with wasting and fasciculation (L.M.N.) as well as hypertonia and hyperreflexia (U.M.N.). This is known as **Tonic Atrophy**.
2. In the lower limbs there will be weakness with signs of U.M.N. lesion. There may be minimal fasciculations if the lumbar A.H.C. are affected.

Treatment of M.N.D.:

1. Avoid fatigue and exhaustion.
2. Massive doses of vit. E and B.
3. Anabolics.
4. Massage and mild exercise.

N.B.: The commonest causes of pseudo-bulbar palsy are:

1. Vascular: – Bilateral stroke (double hemiplegia).
2. Inflammatory: – Encephalitis. – General paralysis of the insane (G.P.I.)
3. Neoplastic: – Midline brain stem tumours. – Syringobulbia.
4. Demyelinating: – D.S.
5. Degenerative: – Motor neurone disease.

N.B.: The commonest causes of true bulbar palsy are:

1. Vascular: – Vertebrobasilar insufficiency.
2. Inflammatory: – Diphtheria. – Bulbar poliomyelitis
3. Neoplastic: – Brain stem tumours.
4. Degenerative: – Motor neurone disease.

D.D. WASTING OF THE SMALL MUSCLES OF THE HAND

The small muscles of the hand are supplied by C_8 and Th1 spinal segments. Wasting of these muscles may be due to:

I. A.H.C. LESIONS:

1) Poliomyelitis:
 - Age: 6 months to 2 years.
 - Acute onset and regressive or stationary course.
 - The wasting is asymmetrical, affecting L.L. more than U.L.
 - No sensory changes.
2) Transverse myelitis: at C_8 and Th_1 segments.
3) Anterior spinal artery occlusion:
 - Acute onset and regressive or stationary course.
 - Both U.L. show flaccid paralysis.
 - Both L.L. show spastic paralysis.
 - There is dissociated sensory loss (touch & deep sensations are intact).

4) Motor neurone disease:
 - Gradual onset and progressive course.
 - Fasciculations in the limbs.
 - Tonic atrophy in the U.L.
 - No sensory changes.

5) Intramedullary lesions:
 - Syringomyelia. – Lower cervical tumours.

II. ANTERIOR ROOTS AND SPINAL NERVE LESIONS:

1) Cervical spondylosis. 2) Cervical Pott's disease.
3) Primary and metastatic tumours of the cervical vertebrae.
4) Fracture and dislocation of the cervical vertebrae.
5) Cervical neurofibromatosis. 6) Pachymeningitis cervicalis.

III. LOWER BRACHIAL PLEXUS LESIONS:

1) Dejerine–Klumpke's paralysis 2ry to birth injury.
2) Thoracic outlet syndrome:
 a) Cervical rib:
 - Only 10% of cases symptomatise.
 - The wasting of the muscles is associated with paraesthesias over the ulnar side of the forearm and hand.
 - The radial pulse is diminished or lost on the affected side when the arm is dragged downwards while the neck is stretched to the opposite side (Adson's test).
 - X–ray may show a complete or incomplete rib.
 b) Pancoast tumour:
 c) Enlarged cervical lymph nodes.
 d) Aneurysm of the subclavian artery.

IV. PERIPHERAL NERVE LESIONS:

1) All causes of mono, mono multiplex and polyneuropathy.
2) Carpal tunnel syndrome:
 - Due to compression of the median nerve in the tunnel.
 - Mainly in middle–aged women and in pregnancy; it may also be 2ry to myxoedema and acromegaly.
 - The thenar muscles are mainly affected, with wasting and weakness.
 - Pain and paraesthesias especially at night associated with sensory loss over the palmar surface of the fingers.

V. MUSCLE LESIONS:

1) Distal type of Gower myopathy. 2) Myotonia atrophica.

VI. Other Causes:

1) Arthritis of the joints of the hand (rheumatoid).
2) Scleroderma & dermatomyositis. 3) Sudek's atrophy.
4) Volkman's ischaemic contractions. 5) Disuse atrophy in long standing U.M.N.L.

SUBARACHNOID HAEMORRHAGE

DEFINITION: Flooding of the subarachnoid space with blood.

CAUSES:

1. Rupture of an intracranial aneurysm which might be:
 a. congenital b. atherosclerotic c. mycotic (e.g. in S.B.E.)
2. Rupture of an intracranial angiomatous malformation.
3. Blood diseases as: purpura, leukaemia.
4. Severe hypertension as in eclampsia.
5. Head trauma.
6. Haemorrhage in a brain tumour.
7. Wrong administration of anticoagulants.

The commonest cause of subarachnoid haemorrhage (S.A.H.) is rupture of an intracranial aneurysm

N.B.: Any intracranial aneurysm may take on one of the following pathways:

1. It may remain **silent** with no manifestations.
2. It may **compress** neighbouring structures, causing pressure symptoms according to its site:
 a. Internal carotid artery aneurysm in the cavernous sinus → compression of Cr. III, IV, VI & ophthalmic division of Cr. V nerves → ophthalmoplegia, facial pain &/or exophthalmos.
 b. Anterior communicating artery aneurysm → compression of optic nerve or chiasma → visual field defects.
 c. Posterior communicating artery aneurysm → compression of Cr. III before entering the cavernous sinus → Cr. III palsy.
 d. Basilar artery aneurysm → compression of midbrain or pons → cranial nerve palsies & long tract manifestations (e.g. hemiparesis).
3. It may manifest only by an **audible bruit**, heard over the skull.
4. It may **rupture**: 90% of cases of aneurysms first present when they rupture, causing the clinical picture of subarachnoid haemorrhage (S.A.H.). Rupture may be **precipitated** by: strain, stress, severe cough or sexual intercourse.

CLINICAL PICTURE:

* Age: Angiomatous group: 1st & 2nd decades.
 Aneurysmal group: 4th & 5th decades.
* Sex: Slightly more in females.

SYMPTOMS:

1. **Headache:** as the meninges are very sensitive & are irritated by the blood.
 - Character: sudden, severe splitting agonising, accentuated by flexion of the neck.
 - Site: starts occipitally & at the back of the upper neck; later on it becomes generalised.
 - Radiation: pain is usually radiating to the shoulders, the lower back & the lower limbs.

2. Symptoms of meningeal irritation:

- Stiffness of the neck.
- Pain in the neck, lower back & limbs due to irritation of the spinal sensory roots.

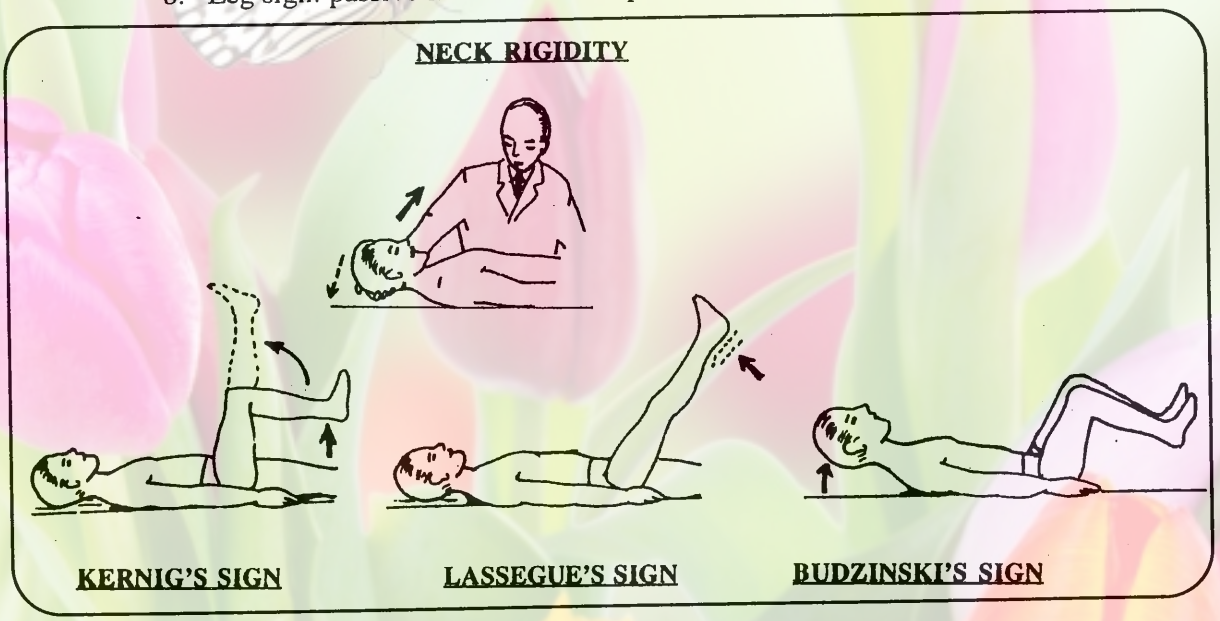
3. General symptoms:

- Nausea, vomiting & vertigo: due to increased intracranial pressure.
- Fever: due to absorption of blood (pyrogenic reaction).

SIGNS:

1) Meningeal irritation signs:

- **Neck rigidity.**
- **Opisthotonus**: high arched back.
- Positive **Kernig's sign**: while the patient is lying in the supine position, with his hip and knee joint flexed at 90°, the doctor attempts to extend the knee. Extension is limited and painful.
- Positive **Lassegue's sign**: while the patient is lying in the supine position with his lower limbs fully extended, the doctor attempts to raise the leg by flexing the hip while the knee is extended. Raising the leg is painful and limited; normally, it can be raised to 90° without causing discomfort.
- Positive **Brudzinski's sign**:
 a. Neck sign: passive flexion of the neck leads to flexion of both knees and hips.
 b. Leg sign: passive flexion of one hip leads to flexion of the other hip and knee.



2) Eye signs:
- Flame–like retinal haemorrhages due to filling of the subarachnoid sheath around the optic nerve with blood.
- Papilloedema, later on, due to increased intracranial pressure.

3) Cranial nerve signs: due to pressure by the blood. The most commonly affected nerves are the ocular nerves (3rd, 4th, 6th) & the optic nerve.

4) Cerebral signs: due to pressure of the blood on surrounding structures:

- Convulsions of different types.
- Hemiparesis or hemiplegia.
- Cortical sensory loss.
- Aphasia.
- Visual field defect.
- Confusion & coma.

INVESTIGATIONS:

1. C.T scan is the first investigation to be done. If blood is detected, the diagnosis is settled and lumbar puncture is not needed.

2. Lumbar puncture for C.S.F. exam:
 - High pressure.
 - Grossly bloody.
 - High protein, but normal sugar and chloride content.
 - Culture is negative.
3. Plain X–ray skull: may show:
 - Calcified wall of aneurysm or angiomatous malformation.
 - Erosion or fracture of bone.
 - Signs of increased I.C.T. (in case of tumour).
4. Angiography: This is an important investigation as it will localise the site of the aneurysm or the angiomatous malformation; it is performed within a few days after the hge.

Differentiation between subarachnoid blood & post–lumbar puncture traumatic blood:

	Subarachnoid haemorrhage	Post-lumbar puncture
1. Blood density	Constant in all samples.	Diminishes in successive samples.
2. Blood clotting	Blood will not clot.	Blood will clot.
3. Supernatant fluid.	Xanthochromia due to disintegration of HB.	Clear

COMPLICATIONS:

1. Rebleeding: its prognosis is worse than that of the initial bleeding.
2. Cerebral ischaemic infarction.
3. Hydrocephalus: due to impairment of C.S.F. drainage by the blood.
4. Epilepsy: due to cortical irritation and damage.

TREATMENT:

1) **Medical**: indicated in:
 1. Acute cases.
 2. Non–surgical cases.
 3. Certain surgical cases e.g.:
 - Multiple aneurysms
 - Huge or inaccessible vascular anomaly.

The medical treatment consists of:

1. Complete rest in bed for 2–3 weeks. Avoid straining, coughing and open the bowels.
2. Analgesics and sedatives to relieve headache.
3. Antifibrinolytic agents: e.g. tranexamic acid and aminocaproic acid; they prevent clot dissolution around the aneurysm and thus prevent rebleeding.
4. Dehydrating measures for the brain.
5. Lumbar puncture for relief of severe headache.
6. Treatment of the cause.

2) **Surgical**: Ligation of the carotid artery or direct surgery to the aneurysm (e.g. clipping of its neck) or angiomatous malformation (e.g. complete excision or occlusion of its feeding vessels). Surgery is best performed 6–14 days after the haemorrhage depending on the patient's clinical condition.

THE CEREBELLUM

ANATOMY: the cerebellum is formed of 2 main parts:

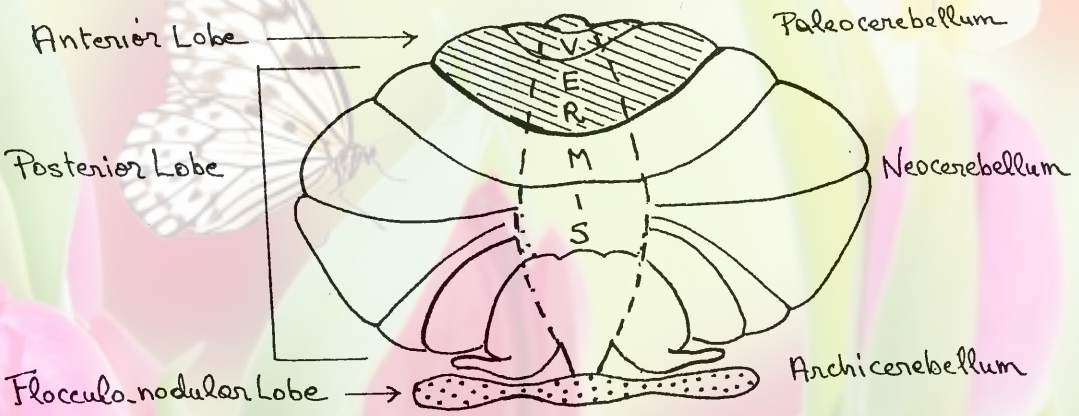
1. A midline central structure, known as the vermis.
2. Two lateral cerebellar hemispheres.

The cerebellum can also be divided in the following manner:

1. Flocculo–nodular lobe. 2. Anterior lobe. 3. Posterior lobe.

HISTOLOGY: the cerebellum is formed of the following:

1. Grey matter forming the cerebellar cortex and the cerebellar nuclei.
2. White matter formed of fibres entering and leaving the cerebellum through the cerebellar peduncles.



PHYLOGENY: phylogenetically the cerebellum can be divided into 3 main parts:

1. **Archicerebellum** (flocculo–nodular lobe) This is the earliest part of the cerebellum to develop. Its main function is the maintenance of equilibrium & it is mainly connected to the vestibular apparatus through the vestibulo–cerebellar and cerebello–vestibular tracts. Lesions of the archicerebellum will lead to **disturbance of equilibrium** during walking or standing.
2. **Paleo–cerebellum** (anterior lobe): This is the next part of the cerebellum to develop. Its main function is the maintenance of muscle tone, it is mainly connected to the spinal cord through the ventral and dorsal spino–cerebellar tracts.
 Lesion of the paleo–cerebellum will lead to disturbance of the muscle tone (**hypotonia**).
3. **Neocerebellum** (posterior lobe): This is the most recent part of the cerebellum to develop and forms most of the bulk of the cerebellar hemispheres. Its main function is the coordination and regulation of the fine delicate voluntary motor activity. It is mainly connected to the cerebral hemispheres through the dentato–rubro–thalamo–cortical tract.
 Lesion of the neo–cerebellum will lead to **incoordination of movements**.

BLOOD SUPPLY: the cerebellum is supplied by:

- Superior cerebellar artery (branch of the basilar artery).
- Anterior inferior cerebellar artery (branch of the basilar artery).
- Posterior inferior cerebellar artery (branch of the vertebral artery).

Cerebellar Syndromes: 2 main syndromes are practically seen:

I. **Archicerebellar syndrome:** There is disturbance of equilibrium of the body manifested by unsteadiness during:
 1. Standing: swaying (trunkal ataxia).
 2. Walking: wide–base or staggering (drunken) gait.

II. **Neo–cerebellar syndrome:** There is incoordination of voluntary motor activities in the form of:
 1. Nystagmus in the eye (fixation nystagmus having rapid and slow components; the rapid component is towards the fixation point while the slow component is towards the resting point).
 2. Dysarthria in the form of staccato speech (explosive, interrupted speech).
 3. Nodding of the head.
 4. Titubation of the trunk.
 5. Intention kinetic tremors in the extremities.
 6. Deviation of the body towards the affected side in unilateral lesions or zigzag gait in bilateral lesions.

N.B.: In both archi–cerebellar and neo–cerebellar syndromes there are:
– Hypotonia and – Hyporeflexia.

ATAXIAS

Definition: Incoordination of voluntary motor activity with or without disequilibration in the absence of motor weakness.

Types: 1. Cerebellar ataxia. 2. Sensory ataxia. 3. Vestibular ataxia.
4. Combined ataxia. 5. Hysterical ataxia.

CEREBELLAR ATAXIA

Causes of Cerebellar Ataxia:

I. **Herido–familial**:
 1. Friedreich's ataxia. 2. Marie's ataxia.

II. **Symptomatic**:
 1. Congenital: – Basilar impression. – Arnold Chiari syndrome.
 2. Infective: – Encephalitis – Cerebellar abscess or tuberculoma.
 3. Vascular: – Superior, middle and inferior cerebellar artery occlusion.
 4. Toxic: – Alcohol. – Barbiturates. – Hydantoins.
 5. Neoplastic: – Medulloblastoma (tumour of the vermis).
 – Astrocytoma (tumour of the hemisphere).
 6. Demyelinating: – D.S. – D.E.M.
 7. Metabolic: – Cortical cerebellar degeneration 2ry to malignancy.

III. **Idiopathic**:
 Delayed cortical cerebellar degeneration in old age

Clinical picture of cerebellar ataxia:

1. **Incoordination** of movements of different muscles in the form of:
 a) Nystagmus in the eyes. b) Staccato speech. c) Nodding of the head.
 d) Titubation of the trunk. e) Kinetic tremors of the limbs.
2. **Hypotonia and hyporeflexia** of the affected muscles.
3. **Gait disturbance**: Wide base or drunken gait in archicerebellar lesions.
4. **Positive tests**: used by the neurologist to detect cerebellar ataxia.

Tests of Cerebellar Ataxia:

1. Finger–to–nose test: The patient brings the tip of his forefinger from a distance onto the tip of his nose. The test is conducted with the eyes open then closed.
2. Finger–to–finger test: The patient brings the tips of his forefingers from the distance of his outstretched arms to meet each other in the midline.
3. Finger–to–doctor's finger test: The patient brings the tip of his forefinger from a distance onto the tip of the doctor's forefinger.
 In any of the above tests you may find:
 a) Decomposition of movement.
 b) Kinetic intention tremors which become more evident as the patient's forefinger approaches the target.
 c) Dysmetria in the form of hypermetria or hypometria.
4. Adiadokokinesis or Dysdiadokokinesis: The patient is asked to do rapidly alternating movements e.g. pronation and supination of the forearm. In cerebellar lesions there is failure to perform the movements.
5. Rebound phenomenon: The patient, with his elbow fixed, flexes it against resistance. When the resistance is suddenly released the patient's forearm flies upwards and may hit his face or shoulder.
6. Buttoning and unbuttoning test: earliest sign.
7. Heel–to–knee test: The patient raises his leg, brings down its heel onto the knee of his other leg & slides it down along the shaft of the tibia.
8. Walking along a straight line, foot close to foot: In unilateral cerebellar lesions there is deviation to the diseased side.


Finger-to-nose test

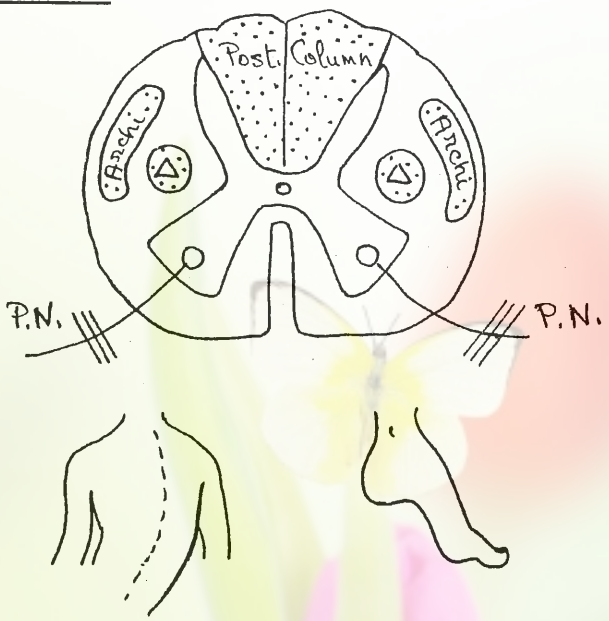

Adiadokokinesis


Rebound phenomenon.


Heel-to-knee test.

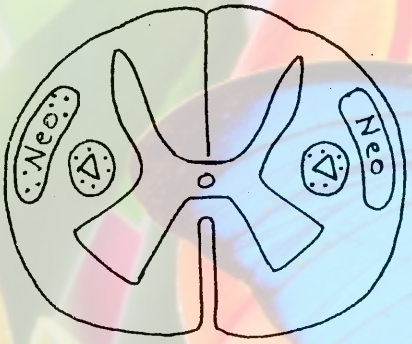
Herido-Familial Ataxias

I. Friedreich's Ataxia



1. It occurs in the 1st decade of life.
2. Gradual onset and slowly progressive course.
3. Positive family history is common as it is inherited as an autosomal recessive disease.
4. Pathologically there is degeneration of:
 - Cerebellum specially the archicerebellum & the spinocerebellar tracts.
 - Pyramidal tracts.
 - Posterior columns.
 - Peripheral nerves.
5. **It presents with:**
 - Progressive cerebellar ataxia of the archicerebellar type i.e. disturbance of equilibrium in the form of gait disturbance and trunkal ataxia.
 - Diminished or lost deep reflexes with positive Babinski sign. The loss of reflexes is due to the degeneration of the peripheral nerves, posterior columns and cerebellum. The positive Babinski sign is due to pyramidal lesion.
 - Impairment of deep sensations (movement, position and vibration), i.e. sensory ataxia due to degeneration of the posterior columns.
 - Stock and glove hyposthesia with lost ankle reflex and preserved knee reflex due to degeneration of the peripheral nerves.
6. Skeletal deformities in the form of pes cavus or kyphoscoliosis are frequent.
7. Congenital heart disease and E.C.G. changes are common.

II. Marie's Ataxia



1. It occurs in the 2nd and 3rd decades of life.
2. Gradual onset and slowly progressive course.
3. Pathologically there is degeneration of:
 - Cerebellum specially the neo–cerebellum (dentate nucleus)
 - Pyramidal tracts.
4. **It presents with:**
 Progressive manifestations of neocerebellar degeneration, i.e. tremors of the extremities, staccato speech, nystagmus . . . associated with exaggerated deep reflexes and positive Babinski sign due to pyramidal tract degeneration.
5. Mental impairment, ocular nerve paralysis and extrapyramidal syndromes are occasionally present.

	Friedreich's ataxia	Marie's ataxia
1. Age of onset	1st decade.	2nd and 3rd decades.
2. Cerebellar manifestations	Mainly archi–cerebellar.	Mainly neo–cerebellar.
3. Deep reflexes	Diminished or lost.	Exaggerated.
4. Sensations	Impaired superficial and deep sensations.	Preserved sensations.
5. Associated findings	Skeletal deformities. Congenital heart disease and E.C.G. changes.	Mental impairment. Ocular nerve palsies. Extrapyramidal syndromes.

SENSORY ATAXIA

DEFINITION: It is ataxia due to loss of the proprioceptive (deep) sensations, at any point in their pathway:

CAUSES:

1. Peripheral nerve: peripheral neuropathy specially diabetic, alcoholic and nutritional.
2. Posterior root: tabes dorsalis.
3. Posterior column: subacute combined degeneration.
4. Medial lemniscus: brain stem lesions.
5. Thalamus: thalamic syndrome.
6. Cortical sensory area: parietal lobe lesions.

CLINICAL PICTURE:

1. Kinetic tremors as tested by finger–to–nose or finger–to–finger tests appear only on **closure** of the eyes.
2. **Rhomberg's test**: when the patient stands with his feet close together & his eyes closed, his body sways & he may fall if not supported.
3. **Stamping gait**: heavy strike of the ground on walking due to lost deep sensation.
4. **Deep sensory loss**.
5. **Hypotonia & hyporeflexia**.



RHOMBERG'S TEST

VESTIBULAR ATAXIA

It is ataxia due to lesions of the vestibular division of the eighth nerve.

CAUSES: 1) Meniere's disease. 2) Labyrinthitis. 3) Acoustic neuroma.

CLINICALLY: There is:

1. Vertigo, tinnitus, deafness.
2. Vestibular nystagmus.
3. Impaired tests for vestibular function.

I CORTICAL LEVEL

II TELENCEPHALIC LEVEL
(Basal Ganglia)

III DIENCEPHALIC LEVEL

IV MESENCEPHALIC LEVEL

V PONTINE LEVEL

VI CEREBELLAR LEVEL



THE EXTRAPYRAMIDAL SYSTEM

THE EXTRAPYRAMIDAL SYSTEM

DEFINITION: It includes all fibres that can influence the motor end plate activity and do not pass in the pyramidal tract.

It is composed of different centres scattered at different levels of the neural axis and all are interconnected via one main cell station (the globus pallidus).

The final influence of this system on the end plate activity is mediated through several descending tracts, mainly the reticulo–spinal tract (page 3).

The different centres and fibres of the extra–pyramidal system are situated at the following levels of the neural axis:

1. Cortical level: mainly in the frontal lobe, and to a lesser extent in the parietal, temporal and occipital lobes.
2. Telencephalic level (**Basal ganglia**): including the **caudate** nucleus and the **lentiform** nucleus (which is formed of the globus pallidus and the putamen).
3. Diencephalic level: including the thalamus, hypothalamus and subthalamus.
4. Mesencephalic level (Midbrain): including the red nucleus and substantia nigra.
5. Pontine level: including the pontine reticular nuclei.
6. Cerebellar level.

Functions of the extrapyramidal system:

1. Regulation and integration of voluntary motor activity.
2. Regulation and maintenance of the muscle tone.
3. Regulation and maintenance of emotional and associative movements.

Disturbance of the functions of the extra–pyramidal system leads to:

1) Disturbance in the integration and regulation of voluntary motor activity resulting in **hyperkinesia** i.e. **STATIC TREMORS** which may be of two main types:
 - Rhythmic and regular as in Parkinsonism.
 - Dysrhythmic and irregular as in chorea, athetosis and dystonia.

 N.B.: These tremors increase with emotional stress, anxiety and fatigue and disappear during sleep and during active voluntary movements.
2) Disturbance in the regulation and maintenance of normal muscle tone resulting in **hypertonia** described as **RIGIDITY**.
3) Disturbance in the regulation and maintenance of emotional and associated movements resulting in **bradykinesia** in the form of **MASK FACE**, infrequent blinking and loss of swinging during walking.

PARKINSONISM

(Shaking Palsy)

It is a condition in which there are static regular rhythmic tremors associated with hypertonia of the muscles of the body and with bradykinesia.

It is due to deficiency of dopamine in the basal ganglia and substantia nigra.

CAUSES:

I. Idiopathic: Parkinson's disease (paralysis agitans). The cause is unknown.
There is degeneration of the pigmented cells (neuromelanin) of the substantia nigra, which becomes pale. The basal ganglia are also affected. This degeneration leads to deficiency of dopamine in the brain.
The age of onset is above 50 years. Both sexes are equally affected.

II. Symptomatic: There is a known cause which leads to deficiency of dopamine in the brain, but without structural changes in the substantia nigra or basal ganglia.
1. Inflammatory: Encephalitis.
2. Vascular: Cerebral atherosclerosis.
3. Toxic: – Co poisoning.
– Mn poisoning.
– Rauwolfia drugs (Reserpine).
– Phenothiazines (Major tranquilisers).
4. Neoplastic: Tumours of the basal ganglia.
5. Traumatic: repeated trauma to the head as in boxers.

CLINICAL PICTURE:

1. **Tremors**:
- Regular, rhythmic and occur at the rate of 4–8/second.
- They begin unilaterally in the U.L. and spread to all 4 limbs.
- They give the hand the **pill–rolling posture** with the thumb moving rhythmically back and forward on the palm.
- They increase with emotional stress and fatigue and disappear during sleep and voluntary movements.



2. **Rigidity**:
- Affecting the proximal more than the distal muscles.
- Affecting more the flexors of the neck, trunk limbs resulting in the **gorilla–like attitude**.
- It may be present throughout the act to the same degree & is then described as lead pipe rigidity; it may be interrupted by the tremors & is then described as cog wheel rigidity.
- Causing difficulty in starting the act of walking leading to a slow, shuffling (**festinant or short steppage**) gait with propulsion.

	Rigidty	Spasticity
Site of lesion	– Extra–pyramidal.	– Pyramidal.
Distribution	– Proximal more than distal. – Flexors of upper & lower limbs and trunk.	– Distal more than proximal. – Flexors of upper limbs, extensors of lower limbs and trunk.
Character	– Lead pipe or cog wheel.	– Clasp knife.
Deep reflexes	– Hyporeflexia.	– Hyperreflexia.

3. **Loss of emotional and associative movements**: resulting in:
 - Immobile face, with frequent blinking (**mask face**).
 - Monotonus speech.
 - Loss of swinging of the arms during walking.
4. **Other clinical manifestations** may be found in some cases (mainly post–encephalitic):
 1) Oculo–gyric crisis: sudden spasm of the conjugate movement of the eyes, mainly upwards.
 2) Greasy face and sialorrhoea. 3) Diabetes insipidus. 4) Obesity.
 5) Impotence or amenorrhoea. 6) Glabellar reflex. 7) Pyramidal signs.

By far the commonest causes of Parkinsonism are:

1) Paralysis agitans. 2) Post–encephalitic. 3) Atherosclerosis.

Features	Paralysis agitans	Postencephalitic	Atherosclerosis
Age	40–60 years	Any age	Over 60 years
Onset	Gradual	Usually acute	Gradual
Course	Progressive.	Regressive or stationary.	Remissions & exacerbations.
Rigidity	Tremors more than rigidity.	Rigidity & tremors equally marked.	Rigidity more than tremors.
Possible associated features		Oculogyric crisis. Greasy face. Sialorrhoea. Diabetes insipidus. Pyramidal signs.	Hypertension. Diabetes mellitus. Pyramidal signs. Coronary heart disease.

TREATMENT OF PARKINSONISM:

I. Medical

The manifestations of Parkinsonism are due to an imbalance between the levels of acetylcholine and dopamine in the basal ganglia and substantia nigra. Reduction of dopamine and relative increase in acetylcholine levels are found in all cases of Parkinsonism, whatever the cause. Therefore medical treatment aims to restore the balance by decreasing acetylcholine or elevating dopamine levels.

1) Anticholinergic drugs:

a. **Natural Belladonna alkaloids** are rarely used nowadays. They include:
– Atropine sulphate. – Hyoscine. – Tincture belladonna.

b. **Synthetic Belladonna like alkaloids**:

* Cogentin 2 mg tab.
* Artane 2 and 5 mg tab.
* Tremaril 5 and 15 mg tab.
* Parkinol 2 and 5 mg tab.

Dose: 1–2 tab. t.d.s., according to the severity of the case.

Side Effects: blurring of vision, dryness of mouth, retention of urine.

Toxic Effects: confusion and hallucinations.

N.B.: Anticholinergic drugs are best prescribed for tremors.

2) Levo–Dopa: (Dopamine precursor):

As dopamine does not cross the blood–brain barrier, its precursor levo–dopa, which can cross it, is used instead.

Dose: $^1/_2$ g orally daily; increase it by $^1/_2$ g every 3 days. Max. dose: 4–6 g daily.

Side Effects:

1) Psychiatric: depression, confusion.
2) Cardiovascular: palpitations, arrhythmias.
3) Gastrointestinal: nausea, vomiting.
4) Neurological: chorea, hypotonia, epilepsy.

3) Levo–Dopa + Carbi–Dopa: (Sinemet)

It was found that L–dopa is to some extent decarboxylated in the liver to dopamine before crossing the B.B.B. resulting in:

– The appearance of side effects due to the peripheral action of dopamine.
– The necessity of using large doses of L–dope to insure the passage of an effective dose across the B.B.B.

These disadvantages are reduced by the addition of **Carbi–dopa** which inhibits the extracerebral decarboxylation of L–dopa to dopamine. Carbi–dopa itself does not cross the B.B.B.

Dose: One tab. of Sinemet contains 250 mg L–dopa + 25 mg Carbi–dopa.

Start with one tab. daily & increase the dose by $^1/_2$ tab. every 3 days till the case improves.

N.B.: Sinemet is best prescribed for rigidity and bradykinesia.

On and off phenomenon: After long term treatment with L–dopa the patient may show signs of Parkinsonism (under treatment) alternating rapidly with signs of drug overdosage, mainly Chorea (overtreatment), as if being turned "on and off." The dose of L–dopa should be reduced and other drugs as Bromocriptine are given.

4) Dopamine agonists: They mimic the action of dopamine at receptor sites.

– Piribedil (Trivastal): 20 mg t.d.s in conjunction with Sinemet.
– Bromocriptine (Parlodel) 2.5 mg daily in conjunction with Sinemet.

5) Amantadine hydrochloride (Symmetrel):

It prevents the uptake of dopamine by the neurones. Dose: 100 mg tab. t.d.s.

II. Surgical

1) Pallidectomy, or
2) Thalamotomy.

CHOREA

DEFINITION: Chorea is involuntary, static, irregular, dysrhythmic, sudden, jerky, pseudopurposive movements of any part of the body, including the face, trunk and/or limbs. It is due to a lesion in the caudate nucleus.

CAUSES:

I. Herido–familial: Huntington's chorea.

II. Symptomatic:

1. Autoimmune: Rheumatic chorea.
2. Infective: post encephalitic chorea.
3. Vascular: Hemiballismus.
4. Toxic: Chorea gravidarum.

III. Idiopathic: Senile chorea.

Rheumatic chorea (Sydenham's Chorea)

It is one of the major criteria of rheumatic fever. It is associated with other rheumatic manifestations in about 10% of cases but never with rheumatic arthritis, and the sedimentation rate is usually normal.

Age: 5–15 years. Sex: females more than males.

CLINICAL PICTURE:

1. **Choreic movements**:
 a) Affecting the tongue, facial, trunk and extremities muscles, being more proximal than distal e.g.:
 - When the patient is asked to keep his tongue protruded and unsupported by his teeth, he is unable to do so, and quickly retracts it.
 - Grimacing, jerking of the shoulders, shaking of the hands and feet.
 b) The movements increase with emotional stress and disappear during sleep.
2. **Hypotonia**:
 - When the patient stretches his arms there is flexion at the wrist and overextension at the metacarpophalangeal and interphalangeal joints with fanning of the fingers giving the boat shaped or scaphoid–shaped hand.
 - When the patient elevates and supinates his arms they deviate downwards and laterally and become pronated.
 - The knee jerk is pendular.
3. **Emotional instability**: As sudden laughter or crying is observed in most cases.

Clinical Varieties:

1. The usual type (described above).
2. **Chorea gravis:** a severe form in which the choreic movements interfere with speech, swallowing and sleep.
3. **Hemichorea:** where the chorea affects one side.
4. **Chorea mollis** (paralytic chorea): due to severe hypotonia, where it may assume a hemiplegic or quadriplegic form with absence of signs of U.M.N. or L.M.N. lesion.
5. **Maniacal chorea:** where there is persistent state of excitement and insomnia.

TREATMENT OF RHEUMATIC CHOREA:

1. Complete rest in bed.
2. Acetyl salicylic acid 6–8 gm daily.
3. Serpasil 1 mg t.d.s. or Haloperidol 3 mg 2–3 times daily.
4. Corticoids as prednisone and prednisolone in cases of:
 - Rheumatic activity.
 - Failure of the above treatment.
5. Adjuvant drugs: tranquilisers and antidepressants.

Chorea Gravidarum

1– Usually occurs in primigravida.
2– It occurs in the 4th and 5th months of pregnancy.
3– It was said that it is secondary to toxaemia of pregnancy but more logically it is rheumatic chorea precipitated by the stress of pregnancy.
4- The condition is treated as rheumatic chorea.

Hemiballismus

1– This is a condition of acute onset in which there are severe violent movements of large amplitude, of one side of he body; these movements may be severe enough to throw the patient off balance or from his bed.
2– It is due to a lesion of the subthalamus (corpus luysi); this lesion is mostly vascular (infarction or haemorrhage) and very rarely due to D.S.
3- Treatment: Phenothiazines or Haloperidol to control the movements.

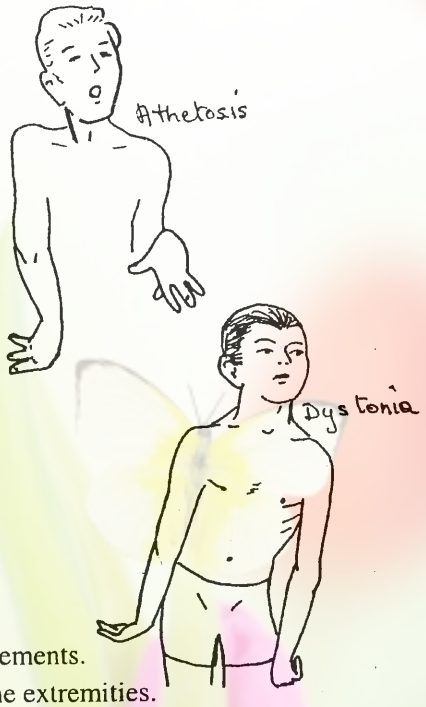
Huntington's Chorea

1– This is a heridofamilial type of chorea inherited as autosomal dominant.
2– It usually occurs in middle age.
3– It has a gradual onset and a progressive course
4– It is due to degeneration of:
 a) The basal ganglia specially the caudate nucleus resulting in choreic movements typically characterised by excessive grimacing and gross movements which interfere with feeding and walking.
 b) The cerebral cortex especially the frontal lobe resulting in marked mentality changes mainly in the form of:
 - Antisocial behaviour with disinterest in people and surroundings.
 - Memory changes.
 - The behavioural and memory changes progress to dementia and the primitive reflexes (grasp, groping, pouting, . . .) can be elicited.

TREATMENT:

1– Genetic counseling: relatives of patients should seek counseling before starting a family, to detect any carrier state.
2- Phenothiazines or Haloperidol to control choreic movements in the early stages.

ATHETOSIS



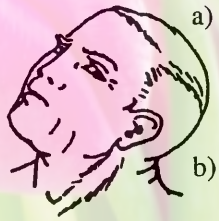
This is a condition where there are:

- Involuntary, irregular, static, slow, snake–like movements.
- They involve the extremities (especially the hands & fingers) and the face on each side.
- They are associated with hypertonia.
- Causes:
 1. Congenital: e.g. Kernicterus & hypoxic neonatal brain damage.
 2. Acquired: Post–encephalitic.
- Treatment: Anticholinergic drugs, the response may be dramatic.

DYSTONIA

This is a condition where there are:

- Involuntary, static, very slow, torsion or twisting like movements.
- They involve the neck, trunk & the proximal muscles of the extremities.
- They are associated with hypertonia during the movement & normal tone in between.
- Dystonia may be:
 1. **Generalised**: **Dystonia muscularum deformans**: it is a familial disorder which starts in childhood. Some cases respond to Levodopa (Sinemet) or Carbamazepine (Tegretol); other cases respond to anticholinergics (e.g. Parkinol).
 2. **Partial**:



a) **Spasmodic torticollis** (wry neck): dystonia of the Sternomastoid leads to turning of the head to the opposite side, specially when the patient is walking; many cases responds to anticholinergics and phenothiazines but some may require myotomy or section of the spinal accessory nerve.

b) **Oromandibular dystonia**: Constant involuntary prolonged tight eye closure (blepharospasm) associated with dystonia of the mouth and jaw muscles.

c) **Writer's cramp**: there is spasm and pain of the muscles of the hand and forearm on attempting to write.

D.D. of Extrapyramidal Irregular Movements

Feature	Chorea	Athetosis	Dystonia
1. Movement	Rapid, jerky pseudopurposive	Slow, snake like.	Very slow, twisting like.
2. Site of movements	Trunk & extremities	Mainly extremities & distal. more than proximal.	Mainly in the trunk & if in the extremities mainly proximal.
3. Tone	Hypotonia	Hypertonia.	Hypertonia during movement and normal inbetween
4. Cause	Commonly, rheumatic fever	Congenital or post–encephalitic.	Familial (in the generalised type).

Tradive Dyskinesia

These are involuntary movements in the face. mouth and tongue (oro–facial) and in the limbs (choreo–athetosis). They are due to long–term treatment with major tranquilisers (phenothiazines e.g. Largactil and butyrophenones e.g. Haloperidol). Response to treatment is poor.

Hepatolenticular Degeneration
(Wilson's Disease)

- It is an autosomal recessive disorder of copper metabolism due to deficiency of ceruloplasmin which normally binds 98% of copper in plasma.
- There is an increase in loosely bound copper–albumin which deposits in all organs.
- The deposition of copper in the basal ganglia causes extra Δ manifestations in the form of choreo–athetosis, dystonia, tremors and/or bradykinesia.
- The deposition of copper in the cornea produces the golden brown "Kayser–Fleicher" ring which is diagnostic.
- Treatment: d–penicillamine, a copper chelating agent is used.



OTHER ABNORMAL MOVEMENTS

1) Myoclonus:

- It is an irregular, asymmetrical shock–like contraction of a single muscle or of a group of muscles. Some cases may be related to epilepsy; others may be related to lesions of the cerebral hemispheres, brain stem, spinal cord or cerebellum.
- **Causes:**
 1. Epileptic disorders associated with myoclonus, e.g.:
 - Aura of grand mal. – Myoclonic petit mal.
 2. Progressive myoclonus:
 - Familial: e.g. lipid storage diseases. – Degenerative: e.g. Alzheimer's disease.
 3. Metabolic disorders associated with myoclonus, e.g.:
 - Hyponatraemia and hypocalcemia.
 - Renal, hepatic, or hypoxic encephalopathy.
- **Treatment:** Clonazepam (Rivotril) + treatment of the cause.

2) Tics: These are involuntary stereotyped repetitive movements (e.g. blinking, shrugging). The patient is aware of them and tries but fails to control them. They occur in neurosis.

3) Fasciculations: Physiological or pathological. See. Neurology Sheet.

4) Static tremors: These are rhythmic oscillatory movements resulting from alternating contractions of opposing muscle groups.

D.D. OF STATIC TREMORS

1. **Parkinsonism.**
2. **Senile**: occur in old age; they are finer, more rapid than in Parkinsonism and are not associated with rigidity.
3. **Essential (familial)**: occur below the age of 25 years; there may be positive family history. They remain stationary throughout life; they respond to Propranolol (Inderal).
4. **Hysterical**: irregular, vary from time to time, associated with other hysterical manifestations.
5. **Hyperthyroidism**: fine, rapid, seen in the outstretched hands and associated with other S & S of thyrotoxicosis.
6. **Hepatic failure**: flapping tremors seen in the outstretched arms; associated with other S & S of hepatic failure.
7. **Toxic**: as in alcohol, mercury and cocaine intoxication.

NYSTAGMUS

Nystagmus is an oscillatory movement of the eyes in a horizontal, vertical, rotatory or mixed direction.

The normal maintenance of ocular posture depends upon:

- Retinal impulses → to the cerebral cortex.
- Labyrinthine impulses → to the brain stem vestibular nuclei and the cerebellum.

In this respect nystagmus may result from:

1. Retinal lesions. 2. Labyrinthine lesions. 3. Brain stem or cerebellar lesions.

I. Retinal or Ocular Nystagmus:

Causes:

1. Physiological: as in case of following moving objects beyond the limits of gaze (optokinetic nystagmus).
2. Pathological: when vision is defective, fixation is impaired and the eyes vainly search; as in cases of congenital cataract and congenital macular defect.

Characters:

* Retinal nystagmus is rapid, pendular & persistent throughout life.

II. Labyrinthine or Vestibular Nystagmus:

Causes:

1. Physiological: as in caloric testing and in rotational acceleration.
2. Pathological: due to lesions of the labyrinth or vestibular nerve.e.g. acute labyrinthitis, Meniere's disease, vestibular neuritis . . .

Characters:

* Vestibular nystagmus has fast and slow phases with the slow phase to the side of the lesion and the fast phase to the normal side. A rotatory component is usually present.
* It is usually associated with vertigo, tinnitus and loss of hearing.

III. Brain stem and Cerebellar nystagmus (Central nystagmus):

Causes:

1. Brain stem lesions e.g. neoplasms, infections, demyelination & vascular lesions.
2. Cerebellar lesions: all causes of cerebellar ataxia.

Characters:

* Central nystagmus has fast & slow phases with the fast phase (direction of nystagmus) to the side of gaze; e.g. if the patient looks to the right the fast phase is to the right and vice versa, so it is "multidirectional."
* In brain stem lesions nystagmus is associated with L.M.N. nuclear cranial nerves paralysis and/or long tracts affection, while in cerebellar lesions nystagmus is associated with other S. & S. of cerebellar ataxia.

N.B.: Dissociated nystagmus (ataxic nystagmus or ophthalmoplegia internuclearis): This is due to lesion of the medial longitudinal bundle in the brain stem wich links the movement of the lateral rectus with the contralateral medial rectus muscle to coordinate horizontal conjugate eye movement.

Clinically when the patient looks to one side there will be nystagmus in one eye (abducting eye) with weaknessof the adducting eye (medial rectus).

BRAIN TUMOURS

Brain tumours are space–occupying lesions within the cranial cavity.

CLASSIFICATION:

1. Tumours arising from the meninges: Meningiomas (arising from the arachnoid).
2. Tumours arising from the brain tissue: Gliomas (astrocytoma, glioblastoma multiformis, medulloblastoma).
3. Tumours of the blood vessels: * Haemangioma.
 * Haemangioblastoma.
4. Tumours of the cranial nerves: Acoustic neuroma of the 8th nerve.
5. Pituitary tumours: * Suprasellar: craniopharyngioma.
 * Intrasellar: acidophil, basophil and chromophobe adenomas.
6. Secondary: * Metastatic from lungs, breast, G.I.T., kidney & prostate.
 * Invasion of the brain by a nasopharyngeal tumour.
7. Congenital tumours: teratoma, cholesteatoma.

CLINICAL PICTURE:

Brain tumours manifest clinically by:

a) General symptoms and signs of increased intracranial tension.
b) Specific symptoms and signs according to the site of the tumour (true localising signs).

A. GENERAL S & S OF INCREASED I.C.T.

The increased I.C.T. is due to:

1. The tumour itself, which is a space–occupying lesion.
2. Impediment of the venous return from the brain.
3. Impediment of the C.S.F. drainage.
4. Haemorrhage into or degeneration of the tumour.

I. Headache:

1. It is due to stretch of the meninges by the space occupying lesion.
2. It is dull, aching, bursting in nature.
3. It is at its peak in the morning and is alleviated as the day passes.
4. It is exaggerated by conditions which increase the intracranial tension as coughing, straining, sneezing and stooping.
5. It has no significant value in the localisation of the site of the tumour; however, generally speaking, supratentorial tumours present with frontal headache, (which is of a late onset) while infra–tentorial tumours present with occipital headache (which is of an early onset).

II. Vomiting:

1. It is due to stimulation of the vomiting centre in the medulla.
2. It is more frequent in the morning.
3. It is not related to meals, and is not preceded by nausea
4. It usually accompanies the headache & may temporarily relieve it.
5. It is more frequent in infra than in supra–tentorial tumours.

III. Papilloedema:

1. This is a reliable sign of increased I.C.T.
2. The patient complains first of blurring of vision followed by gradual diminution of vision: failure of vision follows when post papilloedemic optic atrophy occurs.
3. The field of vision shows concentric contraction.
4. Ophthalmoscopically there is:
 a. Congestion and tortuosity of the retinal veins.
 b. Haziness and blurring of the disc margins.
 c. Filling of the optic cup.
 d. Thinning and attenuation of the retinal arteries.
 e. Retinal haemorrhages and exudates.
 f. Post–papilloedemic optic atrophy may supervene.

IV. False localising signs:

These are signs which may be present regardless of the site of the tumour & are due to → I.C.T.:

1. Lateral ventricle dilatation: mental confusion.
2. 3rd ventricle dilatation:
 - Bitemporal hemianopia (compression of optic chiasma).
 - Hypopituitarism (compression of pituitary gland).
 - Hypothalamic manifestations (comp–ression of hypothalamus).
3. 4th ventricle dilatation:
 - Vomiting (irritation of vomiting centre).
 - Hypertension, bradycardia (irritation of vasomotor centre).
4. Cranial nerve palsies specially the 6th nerve (**abducent**) as it is very thin & runs a long course in the cranial cavity.



5. Herniation syndromes:
 a. **Tentorial (uncal) herniation**: a supratentorial tumour leads to herniation of the uncus of temporal lobe into the tentorial hiatus resulting in:
 * Compression of the reticular formation in the MB → impairment of consciousness.
 * Compression of the 3rd nerve & its nucleus in the MB → dilated fixed pupil.
 b. **Tonsillar herniation**: a subtentorial tumour leads to herniation of the cerebellar tonsils into the foramen magnum resulting in:
 * Compression of the medulla → respiratory irregularities & impairment of consciousness.
 * Tonsillar impaction in the foramen magnum → neck stiffness & head tilt.

B. TRUE LOCALISING SIGNS

These signs depend on the site of the tumour.

1. FRONTAL LOBE TUMOURS:

A. Destructive lesion: one or more of the following:
 1. Mentality changes.
 2. Progressive contralateral hemiplegia.
 3. Motor aphasia and agraphia (if in the dominant hemisphere).
 4. Paralysis of conjugate movement of the eyes, to the opposite side.
 5. Forced grasp reflex.

B. Irritative lesion:
 Focal or Jacksonian contralateral convulsions.
 N.B.: Tumours of the orbital surface of the frontal lobe manifest by the **Foster–Kennedy syndrome** (ipsilateral primary optic atrophy & contralateral papillaedema).

II. PARIETAL LOBE TUMOURS:

A. Destructive lesions: one or more of the following:
 1. Contralateral cortical sensory loss.
 2. Lower quadrantic homonymous hemianopia.
 3. Apraxia, alexia, jargon's aphasia (if in the dominant hemisphere).

B. Irritative lesion:
 Focal or Jacksonian contralateral sensory fits.

III. TEMPORAL LOBE TUMOURS:

A. Destructive lesion:
 1. Auditory agnosia.
 2. Mentality changes.
 3. Upper quadrantic homonymous hemianopia.
 4. Contralateral motor weakness, in deep lesions.

B. Irritative lesion:
 1. Uncinate fits.
 2. Psychomotor or psychosensory epilepsy.

IV. OCCIPITAL LOBE TUMOURS:

A. Destructive lesion:

1. Contralateral homonymous hemianopia.
2. Visual agnosia.

B. Irritative lesion: unformed visual hallucinations.

V. PITUITARY TUMOURS: They may present with:

A. Hormonal Manifestations:

Chromophobe adenoma → hypopituitarism.

Acidophil adenoma → gigantism or acromegaly.

Basophil adenoma → Cushing's syndrome.

B. Neurological manifestations: 2ry to compression of neighbouring structures.

1. Anteriorly:
 - Compression of the optic chiasma → bitemporal hemianopia.
 - Compression of the optic nerve → 1ry optic atrophy.
 - Compression of the olfactory tract → anosmia.
2. Posteriorly:
 - Compression of the upper brain stem → bilateral pyramidal signs and ophthalmoplegia.
3. Laterally:
 - Compression of the optic tract → homonymous hemianopia.
 - Compression of the cavernous sinus → 3rd 4th & 6th cranial nerve paralysis and loss of sensations over the area of the face supplied by the ophthalmic division of the 5th nerve.
4. Superiorly:
 - Compression of the hypothalamus → hypothalamic manifestation e.g. Diabetes insipidus, hypersomnia, adiposity or autonomic epilepsy.

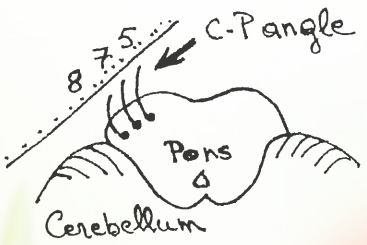
C. Headache: Typically it passes through 3 phases:

- It starts bitemporal due to increased intrasellar pressure.
- Then it disappears due to rupture of the sella turcica.
- Lately it reappears & is generalised with the increased I.C.T.

VI. CEREBELLO–PONTINE ANGLE TUMOURS (C.P.A. tumour): They include:

1. Neurofibroma (acoustic neuroma) of the 8th nerve.
2. Less commonly: • Meningioma. • Cholesteatoma. • Arachnoid cyst.

Clinical Picture of C.P.A. tumour:



1. Ipsilateral cerebellar ataxia.
2. Ipsilateral affection of the 8th, 7th, 5th nerves running in the C–P angle → progressive deafness, facial palsy, facial pain & weakness of the muscles of mastication.
3. Contralateral hemiparesis (Δ compression in pons).

N.B.: The neurofibroma of the 8th nerve may be:

- Solitary.
- Associated with subcutaneous fibromas along peripheral nerves, cutaneous fibroma (mollusca fibrosa) & caf au lait patches in **Von Recklinghausen's disease**.

PATHOLOGY OF BRAIN TUMOURS

I. **Meningioma**: Arises from the arachnoid villi, commonly along the course of intracranial sinuses, it is usually round, encapsulated and compresses the underlying brain tissue without invading it, but erodes the overlying bone with new bone formation from outside (hyperostosis). The tumour may recur after removal.

II. **Glioma**: Arises from the supporting tissues of the C.N.S. and therefore is an infiltrating tumour.

a) Astrocytoma: Most common and most benign. The commonest sites are the cerebral hemispheres in adults and cerebellum and pons in children.

b) Glioblastoma multiformis: Highly malignant and rapidly growing and is fatal within few months. It is liable to haemorrhage and thus may present with an acute apoplectic onset simulating vascular stroke. The commonest sites are the cerebral hemispheres.

c) Medullo–blastoma: Highly malignant, occurring in children with early S. and S. of increased I.C.T. and archicerebellar manifestations. The commonest sites are the roof of the 4th ventricle and vermis of the cerebellum.

III. **Neurofibroma**: Arises from the Schwan cells of the nerve sheaths. It is benign and its commonest site is the 8th nerve in the cerebello–pontine angle (acoustic neuroma).

IV. **Craniopharyngioma**: Arises from the cell–rests of Rathke's pouch, commonly before the age of 15 years. It is liable to calcification and cystic degeneration. It presents by endocrinal manifestations (due to pressure on the pituitary) and by visual disturbances (due to pressure on the optic chiasma, nerve or tract).

INVESTIGATION OF A CASE OF BRAIN TUMOUR

1. Proper history taking and clinical examination.
2. Examination of the field of vision.
3. Ophthalmoscopic examination for papilloedema.
4. Local examination of the skull for:

a) Enlargement. b) Dilated tortuous veins. c) Bony bossses.
d) Angiomatous malformations. e) McEwen's sign: cracked pot note on percussion.

5. **Plain x–ray** of the skull which might show:
 a) General signs of increased intracranial pressure.
 1– Separation of the cranial sutures.
 2– Beaten–silver appearance or finger prints.
 3– Sellar changes:
 – Enlargement of the sella turcica.
 – Rarifaction and destruction of the dorsum sellae and the posterior clinoids.
 – Encroachment on the sphenoid air–sinus
 b) Lateralising signs denoting the side of the tumour:
 Shift of calcified pineal body or falx cerebi.
 c) Signs denoting the site of the tumour:
 1– Localised calcification.
 2– Localised erosion and destruction of the skull bones.
6. **Cerebral angiography**: This may show:
 a) General signs of increased intracranial tension e.g. stretching of all the cerebral vessels.
 b) Shift of the main cerebral vessels (anterior or middle) by the tumour.
 c) Appearance of abnormal vessels feeding the tumour.
7. **Air or myodil ventriculography**: it may show:
 a) Filling defect.
 b) Displacement or deformity of the ventricular system.
8. **Electro–encephalography**:
 Normally the background activity is formed of alpha waves (8–13 c/sec.). In brain tumours there will be asymmetry and disturbance of the background activity either in the form of slow waves in destructive tumours or sharp waves and spikes in irritative tumours.
9. **Echo–encephalography**:
 It may reveal a midline shift, a change in the size of the ventricular system or abnormal echoes due to the presence of the tumour.
10. **C.T. scan**: (computerised tomography of the brain): It shows:
 a. The site and size of the tumour, its density and any cystic degeneration or calcification.
 b. The size of the cerebral ventricles.
 c. Any midline shift.
11. **M.R.I.**: (Magnetic resonance imaging):
 This is the most recent method of investigation. See chapter on D.S.

HYDROCEPHALUS

This is a condition in which there is:
1. increased CSF volume.
2. dilated cerebral ventricles.

CLASSIFICATION & CAUSES:

A. Hydrocephalus with *increased* CSF pressure:

1) **Obstructive**: There is obstruction to the CSF flow.
 a. Non–communicating: the obstruction is within the ventricular system; thus the ventricular system does not communicate with the subarachnoid space. e.g.:
 - Aqueduct stenosis whether congenital or acquired.
 - Intraventricular haematoma, tumour or adhesions.
 b. Communicating: the obstruction is distal to the ventricular system which thus communicates with the subarachnoid space. e.g.:
 - Obliteration of the subarachnoid space by post–infectious adhesion, subarachnoid haemorrhage, or tumours.
 - Occlusion of the venous sinuses by thrombosis or tumours.
2) **Non–obstructive**: there is no obstruction to the CSF flow.
 a. Increased CSF production: choroid plexus papilloma.
 b. Increased CSF viscosity due to increased CSF protein content as in polyneuritis or spinal cord tumour.
 c. Decreased CSF drainage: congenital agenesis of subarachnoid villi.

B. Hydrocephalus with *normal* CSF pressure:

The increased CSF volume & ventricular dilatation are 2ry to atrophy of the brain substance e.g.

a. Congenital cerebral agenesis.
b. Diffuse cerebral atherosclerosis.
c. Dementias as Alzheimer & Hakim–Adam's syndrome.

CLINICAL PICTURE:

a. **Infantile Form**: before closure of skull sutures.
 1. Progressive enlargement of the head with "cracked pot" sound on skull percussion.
 2. Tense anterior fontanelle.
 3. Thin scalp & dilated veins.
 4. "Setting sun" appearance of the eyes due to:
 • lid retraction. • paralysis of upward gaze.
 5. Impaired consciousness & vomiting.
 6. Mental retardation & failure to thrive.
 7. Skull x–ray: separation of sutures & erosion of the posterior clinoids.



b) **Adult Form**: after closure of skull sutures.
 1. S & S of increased I.C.T.: Headache, vomiting, papilloedema, impaired consciousness.
 2. Other S & S may include: dementia, impaired upward gaze, gait ataxia & incontinence.
 3. Skull x–ray: beaten silver appearance & erosion of the posterior clinoids.

TREATMENT

1. Surgical:
 a. Shunt operation to bypass the obstruction & to allow the CSF in the ventricles to drain outside the brain either into the peritoneum (V–P shunt) or into the atrium (V–A shunt).
 b. Ventricular drainage in urgent acutely deteriorating cases.
 c. Removal of a mass or tumour.
2. Medical: reduction of increased I.C.T. using cerebral dehydrating measures as mannitol (or glycerol) & diuretics.

BENIGN INTRACRANIAL HYPERTENSION (B.I.H.) PSEUDOTUMOUR CEREBRI

DEFINITION: This is a condition of increased I.C.T. with no evidence of any intracranial space–occupying lesion, intracranial infection or hydrocephalus (i.e. no → in CSF volume and no ventricular dilatation). The term "benign" is used because there are no serious sequelae except visual impairment.

The increased I.C.T. is due to an increase in the C.S.F. content in the extracellular spaces of the brain leading to interstitial oedema & brain swelling. **Thus the ventricles are not dilated & may even be small.**

CAUSES:

1. **Venous obstruction to C.S.F. absorption** as in:
 - sagittal or lateral sinus thrombosis.
 - CHF.
 - intrathoracic mass lesion.
 - following neck operation.
2. **Idiopathic**: commonest cause occurring mainly in females.
 The mechanism leading to brain oedema is unknown but is related to the following:
 a) Diet: • obesity • hyper/hypovitaminosis A.
 b) Endocrine: • pregnancy • menstrual irregularities • Addisson's disease.
 c) Drugs: • tetracyclines. • steroid withdrawal. • contraceptive pills.
 d) Haematological: • iron deficiency anaemia. • polycythaemia.

CLINICAL PICTURE:

1. Headache is prominent while vomiting is minimal.
2. Blurring of vision due to papilloedema with enlarged blind spot.
3. Transient attacks of loss of vision (amaurosis fugax) denotes a serious threat of complete loss of vision.
4. Diplopia 2ry to 6th nerve palsy (false localising sign).
5. Otherwise, examination of the C.N.S. is normal & the patient looks well.

INVESTIGATIONS:

1. Field of vision: enlarged blind spot: the change in the size of the blind spot is used to monitor the progress of the case.
2. C.T. scan: normal with small ventricles.
3. Lumbar puncture: increased C.S.F. pressure.

TREATMENT:

1. Treatment of the cause, if present e.g. weight reduction diet for obesity, hormonal replacement in Addisson's disease, correction of anaemia, cessation of vitamin A or contraceptive pills intake, . . .
2. Cerebral dehydrating measures:
 - Diuretics: acetazolamide & thiazides (they → C.S.F. production & → diuresis).
 - Steroids: prednisone 20–40 mg/day. After improvement, the dose is gradually tapered to a maintenance dose of 5 mg/day for several months.
 - Mannitol 20% 100 ml infused over 15 min. in acute cases where there is rapidly failing vision prior to surgery.
3. Lumbar puncture: 15–20 ml daily to relieve pressure.
4. Surgical:
 - Lumbo–peritoneal shunt if the above measures fail.
 - Optic nerve decompression to preserve vision in cases of failing vision.

DEMYELINATING DISEASES

1. DISSEMINATED SCLEROSIS D.S.
MULTIPLE SCLEROSIS M.S.

DEFINITION:

This is a disease characterised by affection of mainly the white matter of the C.N.S. resulting in degeneration of its myelin sheaths, and impairment of its function; it is usually of a patchy distribution.

AETIOLOGY:

1) Auto–immune disease as C.S.F. examination shows increase in immunoglobulin content specially immunoglobulin G.
2) Other old theories include: ischaemia of the white matter, viral infection or the presence of an abnormal lipase–fermenting enzyme.

PRECIPITATING FACTORS:

1. Pregnancy and labour.
2. Trauma.
3. Infections as common cold, influenza . . .
4. Physical fatigue.
5. Emotional stress.
6. Surgery.

CLINICAL PICTURE:

a) Age: 3rd and 4th decades.
b) Onset: most commonly acute, but sometimes gradual.
c) Course: most commonly remissions and exacerbations.

d) **Signs and Symptoms**: Any myelinated area of the C.N.S. can be affected resulting in:

1. **Mentality changes**:
 - Euphoria. – Depression. – Emotional lability.
2. **Speech disturbances** (Dysarthria):
 - Slurred. – Staccato. – Scanning.
3. **Cranial nerve involvement** specially:
 - The **optic** nerve resulting in:
 - Diminution or loss of vision due to optic neuritis or primary optic atrophy.
 - Pallor of the optic disc specially on the ***temporal*** side.
 - Visual field defects (specially central scotoma) and disturbances of colour vision due to selective involvement of the macular fibres.
 - The **oculomotor** nerves resulting in:
 - Diplopia which is a common early symptom.
 - Ophthalmoplegia due to weakness or paralysis of the extraocular muscles.
 - ***Ophthalmoplegia–internuclearis*** due to lesion of the medial longitudinal bundle (MLB) which carries the fibres of the conjugate eye movements.
 This sign is pathognomonic of DS in young persons.
 On looking to one side there is:
 a– Nystagmus in the abducting eye.
 b– Weakness or paralysis of the adducting eye.
 - The **facial** nerve resulting in:
 - Facial weakness which may be due to an U.M.N.L. (common) or L.M.N.L. (rare).
 - Hemifacial spasm is rare but if it happens it is pathognomonic of DS.
 - The **cochleo–vestibular** nerve resulting in ***vertigo*** which is a common symptom in DS.
4. **Motor System affection:**
 - This may take the form of monoparesis, paraparesis or less commonly hemiparesis, quadriparesis or pseudo–bulbar palsy.
 - This is associated with signs of U.M.N.L. i.e. hypertonia, hyperreflexia, +ve Babinski sign and ***early loss of the abdominal reflexes***.
5. **Sensory System affection:**
 - Transient numbness and paraesthesias followed by superficial and/or deep sensory loss.
 - ***+ve L'hermite's sign*** may be present: on flexion of the head there is sudden electric like sensation radiating to the back and limbs, it is due to posterior column involvement in the cervical region.

6. **Cerebellar affection:**
 - Cerebellar ataxia is a common presentation; it is associated with nystagmus, staccato speech, intention kinetic tremors and gait disturbances.
7. **Autonomic disturbances:**
 - Precipitancy, hesitancy of autonomic bladder.
 - *Impotence*.

N.B.:

1. Lost abdominal reflexes
2. sphincteric disturbances and } are early findings in DS.
3. impotence.

INVESTIGATIONS:

1. C.S.F. Examinations:

- Cells are increased specially during activity up to 40 cells/mm^3.
- Total protein content is moderately increased.
- Gamma globulin content is usually increased specially IgG.
- **Oligoclonal bands** are found in the CSF in DS.

 In normal CSF, electrophoresis shows a homogeneously stained IgG band; in DS this band shows several distinguishly–stained segments (oligoclonal bands) even if the gamma–globulin content is still within normal.

2. Cortical evoked responses:

The stimulation of any sensory receptor whether visual, auditory or somatosensory evokes an electrical signal in the appropriate region of the cerebral cortex: this is a cortical evoked response. The recording of evoked responses helps in the detection of clinically–unsuspected lesions and the procedure is simple, non–invasive and inexpensive.

- Visual evoked potential (VEP) may show lesion in the visual pathway, which is common in DS.
- Brain stem auditory evoked potential (BAEP) may detect brain stem lesion.
- Somato–sensory evoked response (SSEP) may detect sensory pathway lesion.

3. C.T. Scanning: may show small areas of reduced density corresponding to areas of demyelination.

4. Nuclear magnetic resonance (NMR):

Recently this is the most valuable test. The development of computerised imaging techniques (as in CT scan) has extended its use to identification of hydrogen atom densities in vivo. These directly reflect water content and since this varies from tissue to tissue, NMR can provide a detailed image of intra– and extracranial structures

In DS, NMR shows multiple white matter lesions. Recent and old lesions can be differentiated by the injection of "gadolinium" which appears in the NMR at the site of breakdown of the blood brain barrier in acute lesions.

TREATMENT:

1. Avoid the precipitating factors.
2. Corticosteroids.
 a– A.C.T.H. 80 I.U. by I.M. route dialy for 7 days followed by 40 I.U. (I.M.) for another 7 days.
 b– **Methyl prednisolone 1 gm** by I.V. infusion daily for 7 days recently gives better results.
3. **Beta interferons:** They reduce the frequency and severity of the relapses of D.S., e.g., interferon β 1a (Avonex), interferon β 1b (Rebif) and copolymer (copaxon).
4. Immunosuppressive agents as Azathioprine or Cyclophosphamide are sometimes given but are of doubtful value.
5. A well–balanced diet and high doses of vitamins, specially B1, B6 and B12.
6. Symptomatic treatment.
7. Physiotherapy to maintain mobility and avoid contractures.

II. NEUROMYELITIS OPTICA (DEVIC'S DISEASE)

- It is a type of D.S. which occurs at a young age.
- It is characterised by an acute loss of vision followed by acute paraplegia due to spinal cord lesion. Later on U.M.N. signs settle in.
- The prognosis is poor.

III. BEHCET'S DISEASE

It is an autoimmune disease characterised by:

- Neuropsychiatric manifestations similar to those of D.S.
- Ocular manifestations: hypopyon, Keratitis, iridocyclitis.
- Cutaneous manifestations: buccal and genital ulcers.
- Treatment is as for D.S.

HEADACHE

DEFINITION: Headache denotes pain or discomfort from the level of the brows back to the suboccipital region.

The pain–sensitive structures leading to headache are:

1. The tissue covering the cranium.
2. The dural and cerebral arteries.
3. The intracranial venous sinuses.
4. The meninges, specially the basal meninges.
5. The cranial nerves 5 and 9 and the upper 3 cervical nerves.

N.B.: The pain from supra–tentorial structures is conveyed by the 5th cranial nerve while the pain from infra–tentorial structures is conveyed by the 9th cranial and the upper 3 cervical nerves.

Aetiology:

1. **Vascular headache**: due to vascular dilatation:
 a) Migrainous type.
 b) Non–migrainous type as in systemic infections, hypertension, hypoglcaemia, hypoxic states, vasodilators, temporal arteritis, caffeine withdrawal & alcohol hangover.
2. **Traction headache**: due to stretch of the meninges as in brain tumours, pseudotumour cerebri, intracranial abscess or haematoma and post–lumbar puncture.
3. **Muscle contraction headache** due to prolonged contraction of the muscles of the head and neck resulting in ischaemia with production of metabolites which stimulate pain receptors (e.g. in prolonged driving) & in cervical spondylosis.
4. Headache due to **cranial inflammation** whether extracranial, cranial (cellulitis, osteitis . . .) or intra–cranial (meningitis . . .).
5. **Neuritis and neuralgia** of the sensory nerves of the scalp e.g. trigeminal neuralgia.
6. **Referred headache**: due to diseases of the eyes, ears, nasal sinuses, mouth and teeth.
7. **Psychogenic headaches** (tension headache) in cases of depression and anxiety states; usually in the form of sense of pressure or bandage around the head. This is the commonest cause of headache.



MIGRAINE

It is a paroxysmal often familial disorder characterised by intense throbbing headache (usually unilateral) associated with autonomous manifestations (e.g. nausea & vomiting). It may be preceded by visual, sensory, and/or motor manifestations.

Aetiology:

- Herido–familial (70%).
- Onset around puberty.
- More in obsessive perfectionistic persons.
- Precipitated by mental and physical exhaustion, menses or certain diets e.g. chocolate, cheese, nuts and excessive smoking or alcohol.
- Females more than males (2:1).
- More in urban inhabitants (psychological stress).

Clinical Picture of Classic Migraine:

1) **Prodroma**: drowsiness, fatigue or hunger may occur several days before the attacks.
2) **Aura**: immediately before the attack.
 1. Visual disturbances as scotomatas, flashes of light, zig–zags (fortifications) or hemianopia.
 2. Motor or sensory manifestations; e.g. weakness, aphasia or paraesthesias.

 The manifestations of the aura are present on the opposite side of the coming headache.

 3) **Headache**:

 It occurs in periodic and recurrent attacks. It starts in the temple or around the eye and spreads to involve the whole side of the head. It is throbbing, increases with bright light, excitement and passes away with sleep. It lasts for several hours or days and is associated with nausea, (occasionally vomiting), pallor and coldness of the face and extremities, and may be followed by polyurea.

Mechanism: The manifestations of the aura are due to vasoconstriction of the cerebral arteries while the headache is due to subsequent reactive vasodilatation. There are 2 theories which explain these vascular changes.

1) **Serotinin (5–HT) theory**: An initial increase in blood serotonin level leads to its action on serotonin receptors ($5–HT_1$ receptors) in the smooth muscles of cerebral blood vessels, resulting in their constriction (aura). Degradation of serotonin & decrease of its blood levels follows, resulting in cerebral vasodilatation (headache).
 The serotonin receptors in other parts of the body ($5–HT_2$ & $5–HT_3$ receptors) are similarly affected, resulting in extracerebral vascular changes & activation of some autonomic reflexes (nausea & vomiting).
2) **Ca–uptake theory (hypoxic theory)**: The onset of migraine is due to a focal cerebral hypoxia associated with the rapid entry of Ca into the brain cells & into the smooth muscle cells of the cerebral arteries; this causes the constriction of these arteries (aura) with reactive dilatation (headache).

Important Types:

1. Classic migraine: always preceded by an aura (10% of cases).
2. Common migraine: the headache is not preceded by an aura as the vasoconstrictive phase is not severe. This is the most frequent type of migraine (80% of cases).
3. Ophthalmoplegic migraine: severe migraine followed within days by transient paralysis of the ocular muscles, external and internal ophthalmoplegia.
4. Facial migraine: migraine associated with transient facial paralysis.
5. Hemiplegic migraine: migraine with transient hemiplegia.
6. Basilar artery migraine: it occurs in young females and is often related to menses. The prodroma include visual disturbances, quickly followed by vertigo, ataxia and/or dysarthria. This is followed by severe occipital headache and vomiting.

Cluster headache: (Horton's Syndrome, histamine cephalgia):

1. These are attacks of severe agonising headache starting on one side in the orbital and frontal region, and spreading gradually to the same side of the head and neck. The headache is usually associated with **conjunctival injection, lacrimation, rhinorrhoea**, flushing and sweating of the face on the affected side. There is no nausea or vomiting as seen in other forms of migraine.
2. The attack of headache is brief (several minutes to one hour) and usually occurs during the **night**.
3. 90% of cases occur in **middle–aged males**.
4. The attacks are characterised by their regularity and occurrence in **clusters** i.e. every 24 hours for a few weeks or months. The clusters are followed by long periods (up to 6 months or 1 year), where the patients are completely free.
5. The attacks are precipitated by alcoholic beverages and injection of histamine. They are aggravated by the application of heat and **relieved by cold**.

Treatment of Migraine:

A. During the attack:

1. Ask the patient to try to relax in a dark quiet room; mild attacks may be relieved if the patient can sleep.
2. **Sumatriptan (Imigran)** is a new quick–acting potent drug given during the headache phase of migraine (with or without aura) & in cluster headache.
 - It acts selectively on the serotonin ($5–HT_1$) receptors in the cerebral blood vessels (which are abnormally dilated during the headache phase) leading to their constriction & relief of the headache; the blood vessels in other parts of the body are not affected. Thus Sumatriptan does not have the side effects of Ergotamine which acts on the serotonin receptors all over the body.
 - It acts rapidly within 10–15 min. after S.C. injection.
 & within 30 min. after oral administration.
 - Dose: – 6 mg S.C. may be repeated after 2 hours; max. dose 12 mg/d.
 – 100 mg orally (1 tab.) may be repeated after 2 hours; max. dose 300 mg/d.
 – Nasal spray.
 - It is not used in prophylaxis & like most drugs it is better avoided in hypertension, ischaemic heart disease & in pregnancy.
3. Analgesics as Paracetamol, Optalidon.
4. Antiemetics for vomiting as Metoclopramide (Primperan) in suppository form.

B. Between the attacks:

1. Avoid precipitating factors, if present.
2. Flunarizine HCl (Sibelium): is a Class IV Ca–entry blocker. It selectively prevents Ca overload in the brain cells & in the smooth muscle cells of the cerebral vessels; this protects them from hypoxia responsible for the vascular changes in migraine.
 - Dose: 10 mg (2 caps.) daily before sleep, for 6 months.
 - Side effects: depression, extra Δ disorders, weight gain.
3. Pizotifen (Sandomigran), a serotonin antagonist (one tab. t.d.s.). Start by one tab. daily and add one tab. every 2 days.
4. Propranolol (Inderal), beta blocker; it prevents the uptake of adrenaline by →–receptors in the vessel wall thus preventing vasodilatation. It is best given in common migraine where the vasoconstrictive phase is minimal.
5. Methysergide (Deseril), a serotonin antagonist: 1 mg t.d.s. However, its toxic effects, specially retroperitoneal fibrosis are frequent, which limits its prolonged use.
6. Other measures include:
 - Antihistaminics as Periactin, specially in cluster headache.
 - Antidepressants as Tryptizol, Tranquilisers as Librium, Diuretics as Lasix in premenstrual cases.
 - Antiepileptics as Tegretol if there is a family history of epilepsy or the patient's E.E.G. shows changes suggestive of epilepsy.

TEMPORAL (GIANT CELL) ARTERITIS:

This is a severe arteritis affecting mainly the temporal arteries, but occasionally other arteries of the head (e.g. the ophthalmic artery). It is characterised by:

1) Giant cell infiltration of the media leading to reduction of the lumen of the artery and distal ischaemia.
2) Perivascular inflammation leading to pain and tenderness in the overlying skin.

Clinical Picture:



1) It affects elderly patients (> 60 years).
2) There is severe headache overlying the affected temporal artery.
3) The temporal artery is thickened, prominent, tortuous, non–pulsatile, non–compressible and very tender to pressure.
4) Other symptoms include:
 a– General symptoms: low grade fever, anorexia, weight–loss, sweating, joint and muscle pains.
 b– Visual symptoms specially blindness (unilateral or bilateral) and/or diplopia due to occlusion of the ophthalmic artery.
 c– Jaw claudication i.e. pain when chewing or talking due to ischaemia of the masseter and temporalis muscles.
5) The E.S.R. is always elevated.
6) Temporal artery biopsy shows giant cell infiltration.

Treatment: Prednisolone 30–40 mg/daily for 1 week then reduced gradually over several weeks to 5 mg/daily for several months.

EPILEPSY

DEFINITION:

Epilepsy is a disorder characterised by recurrent, transient attacks of somatic, psychic or autonomic clinical manifestations associated with E.E.G. changes and may be associated with disturbance of consciousness.

AETIOLOGY: (same causes of convulsions).

A. **Idiopathic epilepsy**: i.e. no cause can be found.
- This is the commonest aetiology.
- The family history is positive in some cases.
- It starts in the 1st and 2nd decades.
- It presents with one of the following varieties:
 1. Petit mal epilepsy.
 2. Grand mal epilepsy.
 3. Myoclonic epilepsy.

B. **Symptomatic epilepsy**: i.e. a cause can be detected.

I. Local causes in the brain:
1. Congenital: cerebral palsy.
2. Traumatic: cerebral contusion, laceration or cicatrisation.
3. Inflammatory: – encephalitis. – meningitis. – neuro–syphilis. – brain abscess.
4. Vascular: – hypertensive encephalopathy. – cerebral haemorrhage, thrombosis or embolism.
5. Neoplastic: – primary e.g. meningioma, glioma. – metastatic.
6. Degenerative: – presenile dementias.

II. General causes with 2ry effects on the brain:
1. Toxic: – alcohol, CO, lead, botulism, cyanide, tetanus.
2. Metabolic: – hypoglycaemia. – hyperglycaemia. – hypercalcaemia. – hyponatremia. – Uraemia. – cholaemia.
3. Endocrinal: – hypoparathyroidism. – thyrotoxic crisis.
4. Cardiovascular: – Adam Stoke's attacks and carotid sinus syndrome. – Fallot's tetralogy.
5. Nutritional: – pellagra. – beri–beri. – GABA deficiency.
6. Physical: – high fevers specially in children. – Sun stroke. – Water intoxication (overhydration)
7. Iatrogenic: Metrazol, Ambilhar, Amphetamines, improper use of antiepileptics, E.C.T.
8. Hysterical.

CLASSIFICATION: According to clinical and EEG characteristics.

I- Partial seizures:

They start focally in one region of the brain.

A. **Simple** partial seizures where consciousness is maintained.

B. **Complex** partial seizures where consciousness is impaired.

C. Partial seizures progressing to generalised tonic- clonic convulsions.

II-Generalised seizures:

They start in both cerebral hemispheres at the same time.

A. Absence (**Petit Mal**) seizures.

1. Typical 2. Atypical.

B. Tonic seizures.
C. Clonic seizures
D. Tonic-clonic seizures } components of (**Grand Mal**) fits.

E. Myoclonic seizures.

F. Atonic (Astatic) seizures.

CLINICAL PICTURE:

I- Partial seizures.

A. Simple partial seizures: where there is no loss of consciousness. According to the site of the excitatory focus in the brain the fits may be:

1. Motor: which may be:

- Focal: There is a movement of part of a limb or of the whole limb.
- Jacksonian: There is a movement involving the muscles of one side of the body.It usually has a focal onset either in the thumb, angle of the mouth or big toe depending on whether the wave of excitation in the motor area spreads from above downwards or vice-versa, in a march course.

2. Sensory: which may be:

- General: In the form of paraesthesias (due to irritation of the cortical sensory area) involving one limb (focal) or one half of the body (Jacksonian).
- Special :
 - Visual hallucinations (irritation of the visual sensory area). They may be formed or unformed.
 - Olfactory and gustatory hallucinations (irritation of the uncus). They usually include unpleasant disgusting smells.
 - Auditory hallucinations (irritation of the auditory sensory area).
 - Depersonalisation, derealisation ,deja-vu or jamais-vu phenomena (irritation of the temporal lobe).

B. Complex partial seizures: where there is disturbed consciousness.

The lesion is in the temporal lobe, The fit starts with an Aura which is followed by Absence, Automatism and Amnesia (Syndrome of 4A).

1. Aura: usually in the form of:
 - Olfactory and gustatory hallucinations.
 - Deja-vu or jamais- vu phenomenon.
 - Emotion of fear or elation.
 - Depersonalisation or derealisation.
2. Absence: the patient appears distant with staring eyes and may not respond to questions.
3. Automatism: There may be champing of teeth, smacking, licking of lips or semipurposeful movements of limbs.The patient may walk and leave the room; he may become violent if prevented.
4. Amnesia for the attack.

C. Partial seizures wether **simple** or **complex** may progress to generalised tonic-clonic seizures.

II- Generalised seizures:

A. Petit Mal Epilepsy (Absence seizures):

1. It starts in childhood and improves at puberty.
2. No aura and no sequelae.
3. The fit may be precipitated by hyperventillation or photic stimulation.
4. The fit may be:
 - Typical: there is sudden loss of consciousness of short duration (few seconds), with cessation of motor activity or speech. There is a blank expression on the face.
 - Atypical: the loss of consciousness may be associated with:
 - High frequency (30 -100 attacks/day); the patient does not fall to the ground but looks dazed and staring (Pyknolepsy).
 - Falling to the ground without warning, from which the patient gets up (Akinetic Seizure).
 - sudden very brief jerky movements of the head and or the upper limbs (myoclonic petit mal).

N.B.: Drug of choice: Valproate (Depakin) or Ethosuximide (Zarontin).

B. Grand Mal Epilepsy:

It is characterised by the presence of 3 stages:

1) Pre-ictal stage (aura):
 - It is a warning sign for a coming attack.
 - It gives an idea about the site of onset of the attack.
 - The aura may be somatic, psychic or autonomic.

2) Ictal stage (seizure):
 - Sudden loss of consciousness: lasting not more than 10 minutes.

- Tonic phase (10 seconds) with the onset of coma, the body is thrown into a tonic state with extension of the limbs, rolling of the eyeballs upwards, cyanosis of the face (due to cessation of respiration), clenching of the teeth with tongue-biting and frothing from the mouth, retraction of the head to one side, and urinary incontinence may occur.
- Clonic phase (1-2 minutes): the whole muscles of the body contract & relax repeatdly & rapidly and respiration is stertorous.

3) Post-ictal stage (sequelae):

The sequelae may include lassitude. headache, sleepiness, confusion or Todd's paralysis.

- Todd's paralysis is paralysis following an epileptic fit; it does not last more than 24 hours and is due to neuronal exhaustion.

N.B.: Drug of choice: Carbamazopine (Tegretol) or Phenytoin (Epanutin).

C. Myoclonic Epilepsy:

1) There is a sudden, brief shock-like involuntary contraction of a muscle or a group of muscles.
2) It may occur in the muscles of the face, palate or extremities.
3) It may occur singly (simple myoclonus) or as a part of the aura in grand mal epilepsy or with myoclonic petit mal epilepsy.
4) Juvenile myoclonic epilepsy:
 - It starts between 12-16 years of age.
 - There are frequent myoclonic jerks upon awakening making activities as hair-Combing and tooth-Brushing difficult.
 - The jerks dissapear later in the morning.
 - Few years later the jerks may be accompanied with generalised tonic-clonic seizures.
 - Life long treatment with Valproate is needed.

N.B. Drug of choice: Valproate (Depakin) or clonazepam (Rivotril).

D. Atonic (Astatic) Seizures :

- They start in childhood.
- The attacks occur without warning and last few seconds.
- There is sudden los of postural tone, and the child may fall to the ground.
- Unlike an akinetic seizure there is usually no loss of consciousness,no dazed expression and the response to treatment is poor.

TREATMENT OF EPILEPSY

A. GENERAL MANAGEMENT:

1. Moderation of the patient's physical activities specially swimming, driving or working near machines or at heights for fear of drowning or injury during a fit.
2. Precipitating factors are avoided as photic stimulation (e.g. watching T.V. in the dark) or hyperventilation (e.g. running a long distance).
3. Alcohol intake is forbidden.
4. A ketogenic diet, inducing acidosis, used to be given. Acidosis raises the threshold of stimulation of the brain cells, while alkalosis lowers it.

B. SPECIFIC TREATMENT:

1. Treatment of the **cause** in symptomatic epilepsy.
2. **Anti-epileptic drugs** (see Table) for at least 2 to 3 years.
 a) Always start with one drug (monotherapy); this has several advantages:
 - One drug, in most cases, controls seizures as well as two drugs.
 - It avoids interaction between antiepileptics.
 - It is less expensive and the instructions are easier for the patient to follow.
 b) If the patient does not respond to one drug another drug may be added (add - on therapy) . the new antiepileptic drugs are mainly used as add -on treatment in cases of refractory epilepsy not responding to classical treatment .
 c) Antiepileptics are discontinued only when the patient has been free from fits for at least 2years and his E.E.G. is normal.
 d) Side-effects of antiepileptics include: drowsiness, ataxia, skin rash and blood dyscrasias ; therefore frequent blood pictures should be done. Depakin rarely causes hepatic insufficiency; thus frequent liver function tests are needed early in the treatment . Epanutin also produces gum hyperplasia and hirsutism.
 e) Antiepileptics have teratogenic effect thus an epileptic woman should postpone pregnancy until she is fit-free and off medication; however, if a child is wanted, or if she is already pregnant, she should continue her medication, as the fits are dangerous to her and to her foetus.

ANTIEPILEPTIC DRUGS

Drug group	Dose	Best indicated
1. Barbitarates : **Luminal** (phenobarbitone)	100-600 mg daily	Broad spectrum anticonvulsant NOT for petit mal.
2. Hydontion: **Epanutin**	200-600 mg daily	Simple partial motor seiz. Grand mal // .
3. Carbamazepine: **Tegretol**	400-800 mg daily	Simple partial motor seiz. Complex partial motor seiz. Grand mal seiz. NOT for petit mal.
4. Clonazepam : **Rivotril**	2-6 mg daily	Myoclonic seiz. Grand mal // .
5. Valproate : **Depakin**	600-1500 mg daily	Simple partial motor seiz. Complex partial motor seiz. Petit mal seiz. Grand mal // . Myoclonic // .
6. Succinimide : **Zarontin**	500-1000 mg daily	Petit mal.
7. New Antiepileptics: - Gabapentin (**Neurontin**). - Vigabation (**Sabril**). - Topiramate (**Topamax**). - Lamotrigine (**Lamictal**). - Tiagabine (**Gabitril**).	 1200-2400 mg daily 2000-3000 // // 400-800 // // 200-600 // // 30-60 // //	 Refractory partial and generalised seiz. // // // //. // // // //. // // // //. Refractory Complex partial seiz.

STATUS EPILEPTICUS

This is a medical emergency where there are repeated attacks of generalised convulsions inbetween which the patient does not regain consciousness. The commonest cause is inappropriate withdrawal of antiepileptic drugs. If the convulsions are not stopped rapidly, coma deepens and death may occur due to heart failure, hyperpyrexia or respiratory arrest.

MANAGEMENT:

I. General Management:

1. Guard against the patient falling from the bed.
2. Mouth–gag and O_2 inhalation (endo–tracheal intubation may be needed).
3. Record vital signs regularly; prevent hyperthermia (by sponging.. etc.).
4. Take a sample of:
 - Venous blood for the level of antiepileptic drugs, glucose, electrolytes & C.B.C.
 - Arterial blood for state of pH, pO_2, pCO_2 and bicarbonate level.
5. Start an I.V. infusion of 500 cc normal saline.
6. Cerebral dehydrating measures as Dexamethazone 10 mg I.M., Lasix I.V., concentrated glucose or mannitol 25% I.V.

II. Specific Treatment: Epanutin with Valium (or Rivotril) are given immediately.

1. **EPANUTIN** (Phenytoin): is long–acting and does not depress the respiratory centre.
 Dose: 15 mg/kg slow infusion at the rate of 50 ml/min.
 It is contraindicated in cardiac arrhythmias and in recent myocardial infarction.
 Monitor with E.C.G. and if arrhythmias occur, slow the rate of infusion.
2. **VALIUM** (Diazepam): is rapid & short acting; however, high doses may depress the respiratory center.
 Dose: 5 mg slowly by direct I.V. route; to be repeated after 5 min if seizures recur. maximum dose: 20 mg **OR**
 RIVOTRIL (Clonazepam): Dose: 2 mg directly I.V. every 5 min. max dose; 6 mg.
3. **PHENOBARBITONE** if seizures persist after 20 min of Epanutin & valium administration.
 Dose: 200 mg infusion at a rate of 50 ml/min; respiration should be monitored.
4. In resistant cases **GENERAL ANAESTHESIA** may be used.

N.B.: HYSTERICAL EPILEPSY: (pseudoconvulsions):

* The cause is psychological and there is no organic lesion.
* The fit usually occurs in the presence of other people, to gain their sympathy.
* It never occurs during sleep.
* The patient never hurts himself: there is no tongue–biting or any other injuries to the body.
* Urinary incontinence never occurs.
* The E.E.G. is normal.
* Antiepileptic drugs are contra–indicated.

MENINGITIS

It is the inflammation of the membranes covering the C.N.S., including the dura, arachnoid & pia maters. Inflammation of the dura mater is rare & is known as pachymeningitis. Inflammation of the pia–arachnoid is more common and is known as leptomeningitis.

CLASSIFICATION:

Leptomeningitis can be classified into two groups:

1. Acute pyogenic (purulent) meningitis: The C.S.F. contains mainly polymorphs, due to infection of the meninges by pyogenic organisms as:

1. Meningococci (most common)
2. Streptococci
3. Pneumococci
4. Haemophilus influenza

2. Subacute lymphocytic meningitis: The C.S.F. contains mainly lymphocytes due to infection of the meninges by organisms that do not form pus as:

a) Viruses: Acute anterior poliomyelitis, acute lymphocytic choriomeningitis, mumps and viral encephalitis.
b) Bacteria: Tuberculous bacilli, spirochaetes (as syphilis) and trypanosomes.
c) Fungi: Cryptococcosis and mucormycosis.

MODE OF INFECTION:

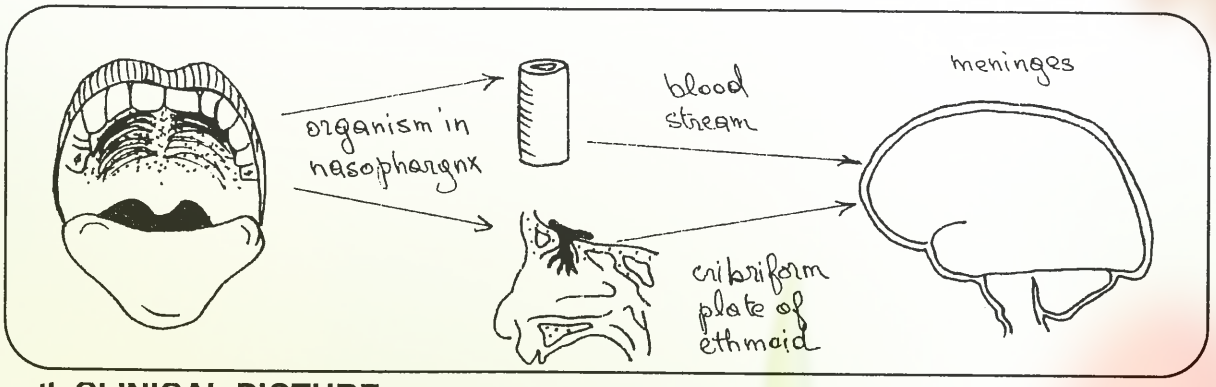
1. Blood stream as in the course of septicaemia or as a metastases from infection of the heart, lungs or other viscera.
2. Direct spread from:
 a) Septic focus in the skull (sinusitis, otitis media), spine (osteitis, Pott's disease) or parenchyma of the C.N.S. (brain abscess, encephalitis, myelitis).
 b) Trauma: organisms may be introduced from outside the C.N.S.(lumbar puncture, penetrating wound) or from the inside (closed head injuries involving the nasal sinuses or the petrous part of the temporal bone).

MENINGOCOCCAL MENINGITIS

(Acute Cerebro–Spinal Fever)

1. AETIOLOGY:

1. The causative organism is the meningococcus which is responsible for 25% of all cases of purulent meningitis.
2. The disease occurs sporadically or in epidemics.
3. The disease spreads by droplet infection and is predisposed to by overcrowding (e.g., schools, soldier's barracks).
4. The disease affects mainly children & young adults.
5. The organisms are implanted on the nasopharynx and gain access to the meninges mainly by the blood stream and to a lesser extent directly through the cribriform plate of the ethmoid bone.



II. CLINICAL PICTURE:

A. Symptoms:

Acute onset with sudden chills, fever, headache, vomiting, backache, blurring of vision, stiffness of the neck and prostration.

B. Signs:

1) **General**:
 1. Temperature: 38°–39°C. may be more.
 2. Pulse is usually rapid except if there is → I.C.T. tension where it will be slow.
 3. Respiratory rate is increased & may be irregular.
 4. Blood pressure is usually normal except in severe cases where there is hypotension.
 5. Haemorrhagic skin rash may appear on the trunk and extremities.
 6. Restlessness and irritability.
2) **Signs of Meningeal Irritation**:
 1. Neck rigidity: passive flexion of the neck is difficult and painful.
 2. Neck retraction and may be opisthotonus.
 3. Positive Kerning, Brudzinski and Lassegue signs.(see p. 99).
3) **Signs of Neurological Deficits**:
 1. Clouding of consciousness, stupor or coma.
 2. Convulsions.
 3. Blurring of the margins of the optic discs.
 4. Transient cranial nerve palsies due to exudation around the nerves.
 5. Diminution of the deep reflexes may occur but other focal neurological signs are rare.

N.B.: Waterhouse-Freidreichson's syndrome is an acute fulminating meningococcal septicaemia associated with:

a. Purpura fulminans causing petechial haemorrhages which may coalesce leading to gangrene.
b. Adrenal haemorrhage (Addisson's crisis) with prostration, low B.P.& coma.

Death may occur within 24 hours due to disseminated intravascular coagulopathy (D.I.C.)

III. COMPLICATIONS:

1) Neurological: – Hydrocephalus. – Deafness.
 – Other focal neurological lesions are rare.
2) Cardiac: – Pericarditis. – Endocarditis.
3) Eye: – Conjunctivitis. – Keratitis. – Iridocyclitis.
4) Genitourinary. – Nephritis. – Epididymitis.
 – Pyelitis. – Orchitis.
5) Joint: Purulent arthritis specially in the knee and shoulder.

IV. INVESTIGATIONS

1) C.T. Scan: to exclude subarachnoid hge. or an intracranial mass.
2) C.S.F. Examination:
 1. The pressure is raised.
 2. The C.S.F. is purulent.
 3. Cells, mainly polymorphs are markedly increased up to 2000–20.000//mm^3.
 4. Proteins are markedly increased. (N: 20–40 mg/dl).
 5. Sugar is markedly decreased to below 20 mg/dl. (N: 50–80 mg/dl).
 6. Chlorides are moderately decreased. (N: 720–750 mg/dl).
 7. Organisms can be detected by **Gram–stain** or **culture** of C.S.F.

Gram –ve intra & extracellular cocci:	Gram +ve diplococci:	Gram –ve bacilli:
Meningococci.	**Pneumococci.**	**Haemophilus.**
		

3) Blood examination for:
 – Leucocytosis up to 30.000/cubic mm.
 – Culture shows the organism in most cases.
4) Detection of the source of infection:
 – Chest X–ray (for pneumonia).
 – Skull X–ray (for a fracture).
 – Sinus X–ray (for sinusitis).

V. TREATMENT:

A) Prophylactic:

1. Isolate the patient & avoid overcrowding during epidemics.
2. Chemoprophylaxis: Rifampicin 600 mg twice daily orally for 2 days for contacts.
3. Immunoprophylaxis: meningococcal live attenuated vaccine $^1/_2$ ml s.c. for children.

B) Curative:

1. General care: proper nursing, adequate fluids, analgesics & sedatives.
2. Specific: once meningitis is suspected start treatment immediately, even before culture:
 a. Antibacterial:
 - Cefotaxime (**Claforan**) 2–4 gm I.V./day, or
 - Penicillin G 20 million units I.V./day or ampicillin 100 mg/kg I.V./day, or
 - Chloramphenicol 100 mg/kg/day in penicillin–sensitive patients.
 b. Corticosteroids: for severe cases as Waterhouse–Freidereichson syndrome along with blood transfusion, dopamine & heparin in suspected D.I.C.

VI. DIFFERENTIAL DIAGNOSIS:

1) Acute general infections: associated with delirium and headache us typhoid, pneumonia and typhus. They are differentiated by the characteristic clinical picture of the disease and by the absence of signs of meningeal irritation.

2) Meningism: This is a condition in which there are signs of meningeal irritation in the absence of meningitis. It occurs in acute infections of children and young adults as typhoid, pneumonia and acute exanthemata. In these cases there is water retention and blood dilution. As the blood is hypotonic to the C.S.F. an attempt is made by the body to reestablish an equilibrium by transferring water from the blood to the C.S.F. through the choroid plexus. As a result the C.S.F. becomes diluted (protein and chloride contents become reduced) while its pressure is raised. This rise of C.S.F. pressure will lead to the symptoms and signs of meningeal irritation which are relieved by lumbar puncture or diuresis.

3) Encephalitis of various types, and brain abscesses: may be associated with signs of meningeal irritation but is differentiated by the presence of early and marked signs of cerebral and brain stem lesion and by the normal sugar and chloride contents of the C.S.F.

4) Subarachnoid haemorrhage: there are signs of meningeal irritation and sometimes fever but the onset is more sudden and the C.S.F. is bloody.

5) Other types of meningitis:

a) Other causes of **pyogenic** meningitis: differentiated by C.S.F. culture.

b) Causes of **lymphocytic** meningitis especially:
 •T.B. meningitis. • Acute lymphocytic choriomeningitis.

T.B. MENINGITIS:

- The onset is insidious and usually preceded by prodromal symptoms of malaise, night fever and sweat and loss of weight (symptoms of toxaemia).
- Signs of meningeal irritation are mild but vomiting and headache are pronounced.
- The presence of a primary tuberculous focus in the body and the typical C.S.F. findings are diagnostic.
- C.S.F. findings:
 1. The pressure is raised.
 2. Appearance is cloudy or ground glass.
 3. Cells, mainly lymphocytes are increased up to 500 cells/cubic mm.
 4. Proteins are increased.
 5. Sugar is decreased to 20–40 mg/dl.
 6. Chlorides are markedly decreased and may reach below 600 mg/dl.
 7. No organisms can be demonstrated by the usual staining methods, but may be revealed by the **Ziehl–Nielsen stain**.

Treatment of T.B. meningitis: Antituberculous drugs:

- Streptomycin 1 gm i.m. daily.
- Isonicotinic acid hydrazide INH 300–800 mg orally daily.
- Rifampicin 600 mg orally daily.

Antituberculous drugs should be given for 1–2 years except Streptomycin which is given for 3 months only for fear of nerve deafness.

ACUTE LYMPHOCYTIC CHORIOMENINGITIS:

This is an acute benign viral infection of the meninges, choroid plexus and rarely the brain substance. It is differentiated from meningococcal meningitis by the presence of excess lymphocytes in the C.S.F. and from T.B. meningitis by the acute onset and by the normal sugar and chloride contents. The virus can be isolated from the C.S.F. by special methods.

ENCEPHALITIS

DEFINITION:

Encephalitis is inflammation of the brain substance.

When the spinal cord is involved the term encephalomyelitis is used.

AETIOLOGY:

I. Primary Encephalitis:

Caused by organisms (viruses) which primarily attack the C.N.S. They are of 2 types:

1. Neurotrophic viruses which attack the nerve cells. They include:
 a. Encephalitis lethargica (Von Economo's disease) or Encephalitis type A.
 b. Rabies. c. Poliomyelitis. d. Inclusion body virus.
2. Pantropic viruses which attack the neurone: These viruses are arthropode borne and cause encephalitis type B.

II. Secondary Encephalitis:

Caused by organisms (viruses, bacteria and parasites), which do not primarily attack the C.N.S. but invade the viscera outside the brain where they acquire the capacity to invade the nervous system.

A. Viral: – Post–infectious as mumps, herpes simplex or zoster and exanthemata.
 – Post–vaccinal as rabies, small pox.
B. Bacterial: Typhoid, bacillary dysentery.
C. Parasites: Malaria, Toxoplasmosis.

CLINICAL PICTURE OF ENCEPHALITIS (1ry or 2ry): 3 sets of features are present:

1) S & S of **viral infection**:
 a. General: fever, headache, myalgia . . .
 b. Specific: depending on the causative organism e.g. L.M.N. limb paralysis in poliomyelitis, parotid swelling in mumps . . .
2) S & S of slight **meningeal** involvement → neck stiffness, C.S.F. changes . . .

3) S & S of **parenchymal** involvement: focal or diffuse.
 - Cerebrum: confusion, dysphasia, hemiparesis, involuntary movements & epilepsy.
 - Brain stem: cranial nerve palsies, long tract manifestations.
 - Cerebellum: ataxia, dysarthria.

In any type of encephalitis, all or some of the above features may be present. The clinical picture varies according to the causative organism & to the site of infection.

ENCEPHALITIS LETHARGICA: "Von Economo's Disease"

It is characterised by:

1. Acute onset with influenza–like symptoms followed after few days by somnolence (varying from drowsiness to deep sleep) with inverted sleep rhythm.
2. Ocular paralysis, oculogyric crisis & Argyll–Robertson pupil.
4. Extra Δ signs: chorea, athetosis or dystonia.
5. Δ signs (monoplegia or hemiplegia) & mental manifestations may occur.
6. **Treatment**: There is no specific treatment. Management includes proper nursing, caffeine & ephedrine for somnolence & antibiotics for 2ry infection.

RABIES: "HYDROPHOBIA":

1. It is caused by the rabies virus present in the saliva of infected "rabid" animals, it is transmitted to man through a bite (usually from a dog) or through skin abrasions.
2. After inoculation, the virus travels to the C.N.S. via the sensory & motor nerves.
3. Incubation period: 1–3 months depending on the site & the severity of the bites.
4. **Clinical picture**: it passes through 3 phases:
 - Period of lethargy, drowsiness & anorexia; there is pain & numbness around the bite.
 - Period of restlessness, excitability, twitchings, convulsions, laryngeal & pharyngeal spasms leading to hydrophobia, hallucinations & a profuse flow of saliva.
 - Generalised paralysis, coma & death from respiratory paralysis.
5. Characteristic feature: Negri bodies found in brain biopsy of the rabid animal.
6. **Treatment**:
 1– Repeated flushing & cleaning of the wound with soap & water.
 2– The wound is not sutured & it is infiltrated with rabies immune globulin or antiserum.
 3– Post–exposure immunisation: both passive & active immunisation are given together.
 a) Passive: The human rabies immune globulin (20 IU/kg); up to 50% of the globulin is used to infiltrate the wound; the rest is given I.M. If human gamma globulin is unavailable use equine rabies antiserum (40 IU/kg/I.M.)
 b) Active: The human diploid rabies vaccine is used I.M. on days 0, 3, 7, 14 & 28 after exposure. If this vaccine is unavailable use the duck embryo vaccine in a series of 23 injections during 2 weeks.

HERPES SIMPLEX :

1. There are 2 types of Herpes Simplex Virus (HSV):
 a. Type I responsible for oral & labial rashes & for encephalitis.
 b. Type II responsible for genital & neonatal infection.
2. Encephalitis results from reactivation of a latent virus rather than from a new infection.
3. C.P.:
 - Onset: headache & fever progressing to impairment of consciousness.
 - As the virus attacks the inferior frontal & temporal lobes, there are olfactory & gustatory hallucinations, behavioural disturbances& complex partial seizures.
4. Treatment: antiviral agent Acyclovir (Zovirax) is a recent successful drug.

HERPES ZOSTER :

1. It is caused by the Varicella–Zoster (VZ) virus which is identical to the virus of chicken pox.
2. H. zoster infection is a reactivation of latent (VZ) virus originally acquired in a childhood attack of chicken pox.
3. The reactivation is facilitated by an immune system weakened by:
 – Steroids. – Malignancy. – Hodgkin's disease. – A.I.D.S.
4. The virus invades the dorsal root ganglia of the spinal cord &/or the cranial nerve sensory ganglia (Cr. V Gasserian ganglion or Cr. VII Geniculate ganglion).
5. **Clinical Picture**: This depends on the ganglion involved.
 a. Dorsal root ganglion involvement: There is a vesicular rash, associated with a burning painful sensation along the course of the affected nerve (dermatome distribution). After 1–3 weeks the vesicles crust over & leave irregular pigmentation & scarring of the skin. Motor weakness may occur due to spread of infection to the spinal cord.
 b. Gasserian ganglion involvement: may result in one of 2 syndromes.
 - H. zoster ophthalmicus: affects the ophthalmic branch of the trigeminal nerve with vesicles & pain above the eye, corneal ulceration, ocular muscle weakness & panophthalmitis.
 - Trigeminal neuralgia: affects the maxillary or mandibular branches of the trigeminal nerve with vesicles & pain along their distribution.
 c. Geniculate ganglion involvement: Ramsay–Hunt syndrome: There are vesicles & pain in the external auditory meatus associated with L.M.N. facial palsy on the same side.
6. Complication: **Post–herpetic neuralgia**. The pain may persist for months or years after cure of the vesicles & is sometimes refractory to treatment.
7. Treatment:
 - Boric acid + zinc oxide: locally to the vesicles.
 - Analgesics & antiepileptics (Tegretol 200–400 mg t.d.s.).
 - Antiviral agent: Acyclovir (Zovirax).
 - Injection of novocaine or phenol in the trigeminal ganglion or resection of the posterior root in intractable pain.

NEURO-SYPHILIS

It is due to affection of the nervous system by the spirochaete Treponima pallidum.

It may affect: 1. The meninges. 2. The vessels. 3. The parenchyma.

I. MENINGEAL SYPHILIS:

1) Cerebral:
 - It occurs in 25% of untreated syphilitic cases, 6 months–2 years after the 1ry infection.
 - There is thickening of the meninges of the base and the convexities.
 - Three clinical forms are recognised:
 1. Asymptomatic: revealed only when lumbar puncture is performed.
 2. Aseptic meningitis: fever, rash, malaise, and neck stiffness.
 3. Acute basal meningitis: manifestations of ↑ ICT (headache, vomiting, papilloedema) and cranial nerve palsies (specially 7th and 8th).

2) Spinal: "Pachymeningitis hypertrophica"
 - There is chronic meningitis with thickening of the meninges of the cervical region resulting in compression of the roots and of the cord.
 - The compression of the roots results in radicular pains and paraesthesias in the U.L. as well as wasting of L.M.N. nature of the muscles of the U.L. supplied by the compressed roots.
 - The compression of the cord results in focal quadriplegia or paraplegia with signs of U.M.N.L.

II. VASCULAR SYPHILIS:

- It occurs 5–10 year after the 1ry infection (tertiary stage).
- It is a syphilitic endarteritis of the large cerebral and spinal vessels.
- It is commonly associated with syphilitic aortitis.
- The clinical picture is that of stroke and depends on the vessel occluded.

III. PARENCHYMATOUS SYPHILIS:

1) General Paralysis of the Insane "G.P.I."

- The onset is 15–20 years after the 1ry infection (tertiary stage).
- There is atrophy of all the brain specially the frontal and temporal lobes.
- Clinically there are 2 stages:
 1. Preparalytic stage: Dementia with changes in memory, affect and behaviour.
 2. Paralytic stage: The dementia is associated with:
 - Δ manifestations as weakness or paralysis with signs of U.M.N.L.
 - Extra Δ manifestations as choreoathetosis.
 - Myoclonic epilepsy.
 - Argyll–Robertson pupil is common.

2) **Tabes Dorsalis**:

- The onset is 15–20 years after the 1ry infection (tertiary stage).
- There is degeneration of the posterior roots and the posterior columns resulting in:
 1. Pains which are severe, knife–like, shooting, lancinating followed later on by marked impairment of the superficial and deep sensations.
 2. The deep sensory loss results in sensory ataxia shown by a +ve Rhomberg test.
 3. The lost muscle sense is known as Abadie's sign.
 4. The deep reflexes are lost and the tone is diminished.
 5. Ulcers of the foot and arthropathies or Charcot's joint are common.
 6. Sphincteric disturbances as sensory atonic bladder are frequent.
- Argyll–Robertson pupil is common.

3) **Rare Forms**:

- Erb's syphilitic paraplegia.
- Syphilitic amyotrophy.
- Syphilitic optic atrophy.

INVESTIGATIONS: C.S.F. examination shows:

1. Increased pressure (in meningitis).
2. Lymphocytosis.
3. Increased proteins.
4. Decreased glucose.
5. Serological tests:
 - The **VDRL** test detects non–specific (Reagin) antibodies. When +ve in the C.S.F., it is diagnostic; if –ve, test for:
 - **FTA** (fluorescent treponemal antibody absorption) or **TPI** (treponema pallidum immobilisation) test which detect specific treponema antibodies. These tests are +ve in every case of neurosyphilis.

TREATMENT:

1. Penicillin G 2–4 million units I.V. 4 hourly for 10 days, or Erythromycin orally for penicillin–sensitive patients.
2. Symptomatic treatment.
3. Physiotherapy in cases of paralysis.

DEMENTIAS

DEFINITION: It is progressive deterioration of intellect (thinking and judgement), memory, behaviour and personality due to disease of the cerebral hemispheres.

CAUSES:

I. Dementia associated with systemic disease:
- Hypothyroidism (cretinism).
- Hypoparathyroidism.
- Cushing's syndrome.
- Liver failure.
- Drug abuse.
- Deficiency diseases (vit. B_1, B_{12}, niacin).

II. Dementia associated with neurological disease:
- Multi–infarction.
- Tumours: e.g.: frontal lobe, temporal lobes, and C. callosum.
- Encephalitis: e.g.: due to Herpes simplex and syphilis.
- Degenerative: Huntington's chorea.
- Normal pressure hydrocephalus.

III. Idiopathic:
- Alzheimer's disease.
- Pick's disease.

CLINICAL PICTURE:

1. Dementia may occur at any age, but is more common in the elderly.
2. The rate of progression depends on the cause. Alzheimer's disease progresses slowly over years, while dementia 2ry to encephalitis may be rapid over weeks.
3. The patient finds increasing difficulty in performing his usual work and social activities. At first he is aware of his disability then loses this awareness.
 Behavioural and personality changes with loss of initiation follow but the patient denies any abnormality.
 In late stages the patient cannot be left unattended and the case ends with mutism and incontinence.
4. The primitive reflexes which are normally inhibited and absent may reappear e.g. grasp, Groping, Glabellar, Pouting and Palmo–mental reflexes.

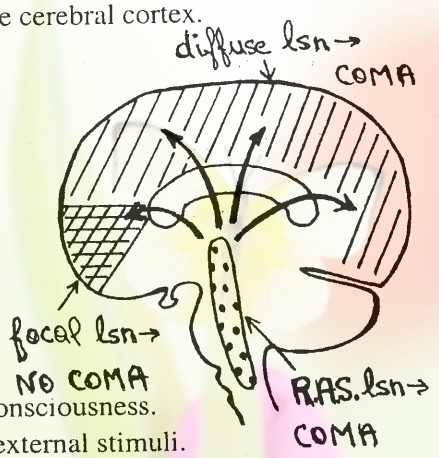
 Grasp reflex: when an object touches the palm of the hand, the hand closes on it (grasps).
 Groping reflex: when an object is moved in front of the eyes, the hand reaches out (gropes) for it.
 Glabellar reflex: (see p. 160).
 Pouting reflex: tapping the lips with a hammer results in a pout response.
 Palmo–mental reflex: a quick scratch on the palm of the hand results in a sudden contraction of the mentalis muscle in the chin.

COMA

INTRODUCTION:

The state of consciousness or alertness depends on:

1. An **intact reticular activating system (R.A.S.)**: This is a collection of nuclei present in the brain stem, hypothalamus & thalamus. It receives impulses from the pathways carrying sensations from the outside world & transmits them through ascending fibres to the cerebral cortex. Its function is the activation of the cerebral cortex.
2. An **intact cerebral cortex**.



Interruption of the state of consciousness may occur at one or both these levels:

a. R.A.S.: a small lesion is sufficient to produce coma.
b. Cerebral cortex: an extensive lesion is necessary to produce coma.

DEFINITION & CLASSIFICATION OF COMA:

1) **Old classification**: based on:
- degree of disturbance of consciousness.
- response of the patient to external stimuli.

State	Consciousness	Response to external stimuli
1. Lethargy or drowsiness	Impaired	Verbal response to increased verbal stimulation.
2. Stupor	Impaired	Verbal response only to vigorous & continuous stimulation.
3. Semi–coma	Lost	No verbal response, only reflex response to painful stimuli.
4. Coma	Lost	No verbal or reflex response to painful stimuli.

2) **GLASGOW COMA SCALE:**

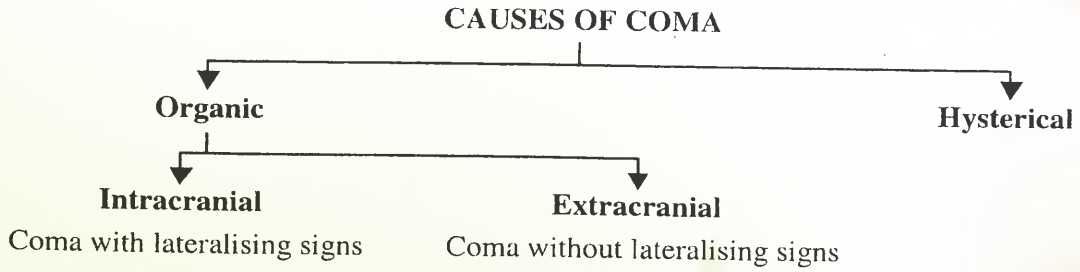
In this scale, the level of consciousness is evaluated, according to the patient's response to external stimuli, using 3 criteria:

- eye opening
- verbal response
- motor response

Each response is given a score. The total score is summed to give an overall value of the level of consciousness from 3–14.

Eye Opening		Verbal Response		Motor response	
Spontaneous	4	Oriented	5	Obeys orders	5
In response to speech	3	Confused	4	Localises to pain	4
In response to pain	2	Words, no sentences	3	Flexes to pain	3
None	1	Sounds, no words	2	Extends to pain	2
		None	1	None	1

According to this scale: coma (score 3) is defined as a state of loss of consciousness where there is no eye opening, no verbal or motor response to external stimuli.



I. INTRACRANIAL CAUSES: the coma is associated with signs of lateralisation:

1) **Traumatic:** e.g. head injury with cerebral concussion, contusion, laceration.

2) **Inflammatory:**
 - Encephalitis: acute onset, fever, headache, confusion, focal cerebral signs.
 - Meningitis: fever, headache, neck stiffness, +ve Kernig's sign, C.S.F. changes.
 - Cerebral abscess: gradual onset of low grade fever, signs of increased intracranial tension, focal cerebral signs, septic focus (e.g. otitis media).

3) **Vascular:**
 - Subarachnoid haemorrhage: sudden onset, severe headache, neck stiffness, +ve Kernig's sign, bloody C.S.F.
 - Cerebral haemorrhage: sudden onset, vomiting, focal signs (e.g. hemiplegia or fits) deepening coma.
 - Cerebral thrombosis: e.g. Basilar artery occlusion: deep coma, decerebrate rigidity, respiratory embarrassment.
 - Cerebral embolism: acute onset, source of emboli e.g. mitral stenosis, A.F., bacterial endocarditis . . ., focal signs as hemiplegia.
 - Hypertensive encephalopathy: hypertension, retinal changes, convulsions.
 - Subdural haematoma: usually middle aged and elderly, headache and fluctuation of consciousness precedes the coma, papilloedema and focal signs may be present ± history of head trauma.

4) **Neoplastic**
 - Primary e.g. meningioma or glioma. – Metastatic.
 - Bilateral papilloedema is usually present.

5) **Epilepsy** (postictal state): history of previous fits; absence of focal signs of a cerebral lesion, dilated pupils, and bilateral extensor plantar response.

N.B.: The signs of lateralisation are:

1. Unequal pupils.
2. Deviation of the eyes to one side.
3. Facial asymmetry.
4. Tilting of the head to one side.
5. Unilateral hypo- or hypertonia.
6. Asymmetric deep reflexes.
7. Unilateral + ve Babinski.
8. Unilateral focal or Jacksonian fits.

II. EXTRACRANIAL CAUSES: (general systemic causes): no signs of lateralisation:

1) Toxic:

- Barbiturates: respiratory depression, circulatory failure & subnormal temperature.
- Opiates: pin–point pupil, respiratory depression, slow weak pulse & cold skin.
- Belladonna (atropine): hot flushed dry skin, fever, dilated pupil, delirium.
- Salicylates: hyperventilation, fever, bleeding tendency & dilated pupils.
- Alcohol: characteristic odour, cold skin, increased level in the blood.
- Co poisoning: cherry red skin, no respiratory distress in spite of O_2 lack.
- Tranquilizers & hypnotics.

2) Hypoxic:

- Pulmonary disease.
- CO_2 narcosis.

3) Ischaemic:

- Cardiac arrest.
- Myocardial infarction.
- Cardiac arrhythmias.
- Hypotensive drugs.

4) Metabolic:

- Hypo & hyperglycemia (D.M.).
- Hypo & hyperthermia (heat stroke).
- Uraemia.
- Cholaemia.

5) Endocrinal:

- Hypopituitarism.
- Hypothyroidism.
- Hypo & hyperparathyroidism.
- Addisson's crisis.

6) Fevers:

- Meningitis, encephalitis.
- Malaria specially cerebral type.
- Septicaemia.
- Status typhosus.

CAUSES OF FEBRILE COMA:

1. Infective: encephalitis, meningitis & other hyperpyrexias.
2. Vascular: pontine haemorrhage, subarachnoid haemorrhage.
3. Metabolic: diabetic ketoacidosis, hepatic cirrhosis.
4. Endocrinal: thyrotoxic & Addissonian crisis.
5. Toxic: Belladonna & salicylate poisoning.
6. Sun stroke & heat stroke.
7. Coma with 2ry infection due to hypostatis pneumonia, U.T. infection or bed sores.

CLINICAL APPROACH TO A CASE OF COMA

I. HISTORY: taken from the patient's relatives.

1. Onset: may be:
 - Sudden e.g. cerebral haemorrhage or embolism.
 - Subacute: e.g. cerebral thrombosis.
 - Gradual e.g. brain tumour.
2. Head injury: cerebral concussion, laceration or haemorrhage.
3. Convulsions: post–ictal coma, brain tumour or overdose of antiepileptics.
4. Drug intake: insulin overdose, drug intoxication (e.g. barbiturates . . .).
5. Exposure to the sun as during the Pilgrimage in Mecca (sun stroke).

II. GENERAL EXAMINATION

1) **Temperature**:
 - Hyperthermia: causes of febrile coma (see above).
 - Hypothermia: – hypopituitarism, hypothyroidism.
 – barbiturate, opiate or alcohol poisoning.
 – peripheral circulatory failure: cardiac causes.

2) **Pulse**:
 - Bradycardia: brain tumour, opiate poisoning, myoexedma.
 - Tachycardia: hyperthyroidism, uraemia . . .

3) **Blood pressure**:
 - High B.P.: hypertensive encephalopathy.
 - Low B.P.: Addissonian crisis, alcohol & barbiturate poisoning.

4) **Respiration**:
 1. Slow, deep, stertorous: in morphine & barbiturate poisoning.
 2. Rapid, deep (Kussmaul) respiration: in diabetic or uraemic acidosis.
 3. Hyperpnoea regularly alternating with apnoea (Chyne–Stokes respiration): lesions affecting both cerebral hemispheres.
 4. Central neurogenic hyperventilation: similar to Kussmaul's respiration but the cause is a lesion at the junction between midbrain & pons.
 5. Apneustic breathing: prolonged pause at full inspiration due to pontine lesion.
 6. Ataxic breathing: phases of deep & shallow breathing alternate irregularly: due to medullary lesion.

5) **Odour of breath**:
 1. Acetone odour: in diabetic ketosis coma.
 2. Fetor hepaticus: in hepatic coma.
 3. Uriniferous odour: in uraemic coma.
 4. Alcohol odour: in alcohol intoxication.

6) **Inspection of skin**:
 1. Injuries or bruises: in traumatic causes.
 2. Dry skin: in diabetic ketosis, atropine poisoning.
 3. Moist skin: in hypoglycaemic coma.
 4. Cherry–red colour: in CO poisoning.
 5. Needle marks on limbs: in drug addiction.
 6. Rashes: in Waterhouse–Freidreichson's meningitis, in endocarditis & other exanthemata.

III. C. N.S. Examination:

1) **Signs of lateralisation**: denoting an intracranial cause for the coma. (See p 148).
2) **Pupillary signs**:
 1. Dilated, irreactive to light.
 – Unilateral: 3rd nerve compression, as in uncal herniation.
 – Bilateral: e.g. atropine poisoning.
 2. Constricted:
 – Unilateral: Horner's syndrome (p 22); however, alone, this syndrome does not cause coma.
 – Bilateral:
 - Reactive to light: metabolic coma.
 - Irreactive to light: pontine haemorrhage, morphine poisoning (pin–point pupil).
3) **Fundus examination**: for papilloedema in cases of increased I.C.T.
4) **Signs of meningeal irritation**: Neck stiffness, opisthotonus, +ve Kernig's & Lassegue's signs in cases of meningitis & subarachnoid haemorrhage.

IV. Laboratory Investigations:

1) Blood examination:
 1. Blood picture: leucocytosis in bacterial infections.
 2. Blood film for malaria parasites.
 3. Blood levels of sugar, urea, creatinine, bilirubin, . . .
 4. Blood gases: pH, HCO^{-3}.
 5. Toxicological studies in cases of poisoning.
2) Analysis of gastric contents for possibility of poisoning.
3) Urine analysis: for sugar, acetone & albumin.
4) C.S.F. examination: e.g. bloody in S.A.H., purulent in bacterial meningitis . . .
5) Plain x–ray skull: e.g. sellar changes in ↑ I.C.T., fractures in trauma . . .
6) C.T. scan & M.R.I. show the site, size and nature of intracranial lesions.

V. Management of Coma:

A. Treatment of the cause.

B. General care of the comatosed patient:

1– Care of the skin with frequent changing of the patient's posture & washing of the skin of the back & pressure points with alcohol followed by talc powder.

2– Care of respiration with suction of secretions and O_2 inhalation.

3– Care of nutrition and fluid balance with tube feeding and I.V. fluids.

4- Care of the urinary bladder with urinary antiseptics & a catheter if necessary.

5- Care of the bowels with a daily enema.

NEUROLOGICAL SHEET

I. HISTORY

I. PERSONAL HISTORY: Ask about:

1. **Name**: To be familiar with the patient.
2. **Age**: As certain diseases are more common in certain ages e.g.
 1st & 2nd decades: progressive muscular dystrophy.
 3rd & 4th decades: DS.
 5th & 6th decades: Cerebrovascular strokes.
3. **Sex**: Motor neurone disease (M.N.D.) is common in males.
 Migraine is commoner in females.
 - Ask about contraceptive pills as they may cause headache, depression or DVT.
4. **Marital state**: For possible sterility, impotence or still–births (as in syphilis).
5. **Occupation**: Persons in certain occupations are more susceptible to certain diseases e.g. disc prolapse is commoner in drivers while lead neuropathy is commoner in printers.
6. **Residence**: e.g. migraine is commoner in urban areas while nutritional diseases are commoner in rural areas.
7. **Special habits**: e.g. alcohol can lead to peripheral neuropathy.
8. **Handedness**: In right–handed people (over 90% of population), the dominant hemisphere is the left.

II. COMPLAINT:

Put it in the patient's own words & list his complaints according to their importance.

III. PRESENT HISTORY: "Analysis of the complaint:"

It includes Duration, Onset, Course & Sequence of Events in Chronological order e.g.: The condition started since . . . (duration) by acute or gradual (onset) & regressive or progressive (course) of . . . Tell the story of the disease chronologically & in details.

Then ask about the following symptoms if the patient did not mention them

1. Symptoms suggestive of increased Intracranial Tension (↑ICT):

1. Headache. 2. Vomiting. 3. Blurring of vision.

2. Symptoms of Cranial Nerve Affection:

Cranial	Nerve	Symptom of Lesion
I	Olfactory	Anosmia – Parosmia.
II	Optic	↓ Acuity of vision – Field defects
III, IV, VI	Ocular Nerves	Diplopia
V	Trigeminal	Difficult mastication (motor) Abnormal face sensation (sensory)
VII	Facial	Accumulation of food behind cheek.
VIII	Cochleo–vestibular	↓ Acuity of hearing, Tinnitus (cochlear part).
		Vertigo (vestibular part).
IX, X, XI & XII (Bulbar Nerves)	Glossopharyngeal & vagus Cranial accessory & Hypoglossal	– Dysphagia – Dysarthria – Dysphonia (hoarseness of voice) – Nasal regurgitation.

3. Symptoms of Motor System Affection: (U.M.N., L.M.N., extra Δ, cerebellum):

1. Destructive lesion: weakness or paralysis. If present ask about muscle tone & wasting.
2. Irritative lesion: Convulsions, fasciculations and/or abnormal movements.

4. Symptoms of Sensory System Affection:

1. Destructive lesion: Hyposthesia or anaesthesia.
2. Irritative lesion: Pain, hypersthesia and/or parasthesia (abnormal sensations of skin).

5. Symptoms of Autonomic (Sphincteric) Disturbances:

1. Control of micturition & defaecation.
2. Impotence (specially in cases of conus lesions, DS & diabetic PN.).

IV. PAST HISTORY:

1. **Trauma**: Usually severe: in cases of paraplegia, quadriplegia, cauda lesions & coma.
 * Mild trauma to the head might cause subdural haematoma in old alcoholics
2. **Fever**: specially near the onset of the disease. In cases of: Meningitis, encephalitis & myelitis.
3. **Diabetes Mellitus** (polyuria, polydipsia, polyphagia & weight loss): In cases of peripheral neuropathy, cranial N. palsy & impotence.
4. **Hypertension** (headache, tinnitus, epistaxis): In cases of hemiplegia, cerebral haemorrhage, encephalopathy.
5. **T.B.** (haemoptysis, symptoms of toxaemia as night fever, night sweats, loss of weight, appetite & anti–TB drug intake): In cases of paraplegia (Potts), cerebellar ataxia and meningitis.
6. **Syphilis** (chancre, recurrent still–births, abortions): In cases of sensory ataxia (Tabes) mental deterioration with convulsions (G.P.I.).
7. **Rheumatic fever & R.H.D.** (arthritis, epistaxis . . .): In cases of hemiplegia & chorea.

8. **Otitis media** (ear discharge): In cases of facial palsy, brain abscess & lateral sinus thrombosis.
9. **Previous drug intake**: In cases of:
 - Cerebellar ataxia: Barbiturates, Hydantoin.
 - Convulsions: Ambilhar.
 - P.N.: Streptomycin, I.N.H., Sulphonamides.
 - Myopathy: Vincristine, Chloroquine.
 - Parkinsonism: Reserpine, Phenothiazydes & other major tranquilizers.
10. **Previous similar attacks**: In cases of D.S. & T.I.A.s.

V. FAMILY HISTORY: Ask about:

1. Similar conditions in the family.
2. Consanguinity between parents.

II. EXAMINATION

A. GENERAL EXAMINATION:

Before proceeding to the examination of the nervous system conduct a thorough general examination: General appearance, pulse, temperature, blood pressure, heart, chest & abdomen.

B. NEUROLOGICAL EXAMINATION:

1. EXAMINATION OF THE MENTAL FUNCTION

Report on:

1) **State of consciousness**: For assessment (see p. 147).
2) **Orientation for time & place**: Ask: "What time is it? What place is this?"
3) **Memory**:
 - It is the ability to retain & recall informations & experiences.
 - It is mainly the function of the limbic system of the temporal lobe.
 - Test for: a. **Immediate** memory: Tell the patient a group of digits (numbers) & ask him to repeat them. Find out the no. of digits he can repeat correctly after one hearing (e.g. 5, 9, 7, 12 . . .). Normally he should recall 7 digits.
 b. **Recent** memory: Ask the patient if he remembers some recent events e.g. "What did you have for breakfast yesterday?" & check his answers with his surrounding family.
 c. **Remote** memory: Ask the patient if he remembers some old events e.g. "What year was the Egyptian Revolution?"
 - Diminution of memory is termed **AMNESIA** which includes:
 a. **Anterograde** amnesia: loss of memory for immediate & recent events.
 b. **Retrograde** amnesia: loss of memory for remote events.
 c. **Transient** global amnesia (circumscribed amnesia): sudden total loss of memory lasting for less than one day in a middle–aged healthy person, it may be precipitated by physical or emotional stress. It may be due to temporal lobe ischaemia & the condition is benign.

- Commonest **Causes of Amnesia**:
 1. Cerebral atherosclerosis.
 2. Temporal lobe lesions.
 3. Korsakow's syndrome due to chronic alcoholism (amnesia, confabulations & P.N.).
 4. Dementias e.g. Alzheimer & Huntington's chorea.
 5. Post–concussion.
 6. Hysterical.

4) **Mood & Affect**:
 - Mood is the patient's inner **feelings** while
 - Affect is the outward **expression** of emotion.

Abnormalities in Mood and Affect include:

1. Depression.
2. Euphoria.
3. Emotional lability.
4. Apathy or indifference.

The commonest causes of abnormalities of Mood & Affect are:

1. D.S.
2. Pseudo–bulbar palsy.
3. Cerebral atherosclerosis.
4. Psychosis and Neurosis.

5) **Intelligence:**

It is usually assessed by special "Intelligence Quotient" (I.Q.) tests. For simplicity, the patient is considered of average intelligence when he and the doctor can understand each other.

6) **Behaviour:**

It is the overall manner in which the patient sits, dresses, talks and cooperates with the doctor.

In case of a **normal mentality** report as follows:
The patient is fully conscious. well oriented for time and place, with normal memory and mood; he is cooperative and of average intelligence.

N.B.:

- **Hallucination**: is a sensation without an external stimulus. It might be visual, auditory, olfactory or tactile.
- **Illusion**: is misinterpretation of an external stimulus.
- **Delusion**: is a false fixed belief, not correctable by reasoning and not shared by others of the patient's same culture.

 Delirium: is a transient state of acute confusion (disorientation) in which there is restlessness, hyperexcitability and where the patient suffers from illusion and hallucinations. It occurs in acute infective fevers, alcoholism (delirium tremens), certain drug intoxication (belladonna, amphetamine).

2. EXAMINATION OF SPEECH

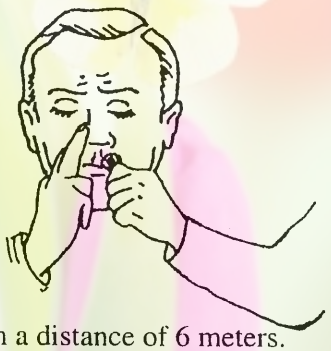
Notice during history taking and comment. The main speech disturbances include:

1. **Aphasia**: Inability to formulate speech. Types:
 1) Sensory aphasia: a) Visual agnosia and alexia. b) Auditory agnosia.
 2) Motor aphasia: a) Verbal aphasia. b) Writing aphasia (agraphia).
2. **Dysarthria**: Difficulty to articulate speech properly.
 1. Staccato speech: in cerebellar lesions.
 2. Slurred speech in pyramidal and L.M.N. lesions of speech muscles.
 3. Monotonous speech in Parkinsonism.

 {See details in chapter on speech, Page (35)}.

3. EXAMINATION OF THE CRANIAL NERVES

1. OLFACTORY NERVE:

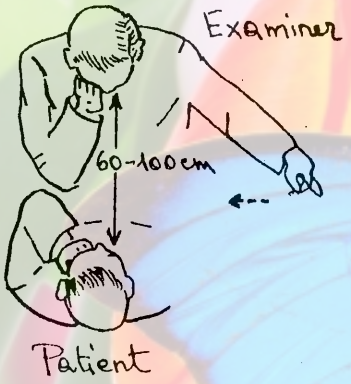


- Examine for the sense of smell using a familiar non–irritant substance (e.g. ground coffee).
- Each nostril is tested separately with the patient's eyes closed.

Anosmia means loss of sense of smell (see p 15).

2. OPTIC NERVE: Examine for:

1. **Acuity of Vision**: Using Snellen's chart or finger counting from a distance of 6 meters.
 In case of failure to count the fingers at this distance repeat at a shorter distance.
 If at a distance of 30 cm the patient still fails to count the fingers test for vision using hand movements.
 If the patient does not see the movements, test for light perception using the torch.
 If there is no P.L. then the patient is blind.
 Each eye should be examined separately.
2. **Field of Vision**:
 It is examined using Bjerrum's screen (central vision) and the perimeter (peripheral vision). If these are not available use the confrontation test.
 - Sit in front of the patient at a distance of 60–100 cm. Keep your eyes at the level of the patient's eyes.
 - Let the patient close one eye and you close the opposite eye. Insist that the patient looks into your eye and nowhere else.
 - Examine for the field of vision of the patient's open eye by bringing your finger slowly from the periphery inwards. Test for the whole field by bringing your finger from above, below, left and right.



N.B.: The patient's field of vision is normal when he notices the movement of your finger at the periphery of the field at the same time that you do. Then test for the other eye in the same manner.

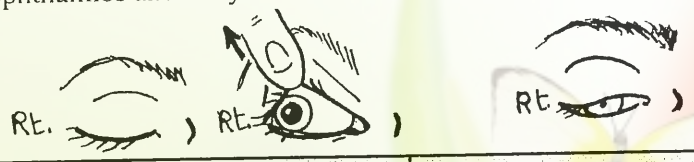
3. **Ophthalmoscopic examination** for papilloedema & optic atrophy (See Brain Tumours).
4. **Colour of vision** for colour blindness.

3, 4, 6, OCULAR NERVES:

Examine for:

1. Ptosis which may be due to:

- Oculomotor nerve paralysis where the ptosis is complete and there is associated mydriasis and divergent squint.
- Sympathetic paralysis (Horner's syndrome) where the ptosis is partial and there is associated miosis, enophthalmos and anhydrosis.



	Third N.	Sympathetic
Degree	Complete	Partial
Pupil	Dilated and fixed (Mydriatic)	Small (Miotic)
Association	Divergent squint	Enophthalmos, anhydrosis

* To determine whether the ptosis is partial or complete, the action of the frontalis muscle should be abolished. Press a finger over the superior orbital margin, then ask the patient to open his eye: if he can, the ptosis is partial while if he cannot, the ptosis is complete.

- **N.B.**: Myasthenia gravis is the commonest cause of ptosis; however the ptosis is usually bilateral, the pupil is normal and there are other myasthenic manifestations.

2. Pupils: They should be equal, round and reactive to light, and accommodation.

- The **light reflex**: If you expose one eye to light, while shading the other, normally there is constriction of the pupil of the exposed eye (direct reaction) as well as of the other eye (consensual reaction).
- The **accommodation** (near) **reflex**: When the patient is asked to follow your finger with both his eyes from a far to a near point, the following triad normally occurs:
 a) Convergence. b) Miosis. c) Accommodation.
- **Cilio–spinal reflex**: Pinching the skin on one side of the neck results in dilatation of the ipsilateral pupil. This reflex is absent in cervical sympathetic lesions (Horner's syndrome).

3. Extra ocular movements:

- Test for the **abducent** nerve (supplying the lateral rectus muscle) by asking the patient to look laterally.
- Test for the **trochlear** nerve (supplying the superior oblique muscle) by asking the patient to look inwards & downwards.
- Test for the **oculomotor** nerve (supplying the superior, medial and inferior rectus and the inferior oblique muscles) by asking the patient to look in all other directions.
 - These tests are done for each eye alone: If their results are normal this indicates that the ocular nerves are intact.
 - Then repeat the same tests on both eyes simultaneously for conjugate movement; if normal, then the centres for conjugate movements present in the brain stem, and in the frontal and occipital lobe cortex are intact.

4. Nystagmus:

Ask the patient to look at your finger placed laterally, upwards then downwards, at some distance from his eyes. If nystagmus is present comment:

- If it is spontaneous or on fixation.
- If it has rapid and slow phases; the direction of the nystagmus is that of the rapid phase.

5. TRIGEMINAL NERVE:

a) Motor Part:

1. Test for the power of the muscles of mastication:
 - **Temporalis**: Ask the patient to clench his jaws while you put your hands over the temples to palpate the muscles.
 - **Masseters**: The patient clenches his jaws while you palpate the contracted muscle between four fingers over its posterior border and the thumb over its anterior border.
 - **Pterygoids**: Ask the patient to open his mouth while you fix his head.
 Unilateral paralysis: Deviation of the jaw to the diseased side.
 Bilateral paralysis: No deviation, but inability to open the mouth against resistance.
2. **Jaw reflex**: (afferent Cr. 5 efferent Cr. 5)
 While the mouth is slightly open, place your index finger on the lower jaw, and then tap it from above downwards.
 * Normally the reflex is absent or minimal.
 * An exaggerated reflex, shown by closure of the jaws, denotes a bilateral U.M.N.L. above the motor nucleus of the 5th cranial nerve i.e., above the pons as in pseudobulbar palsy.

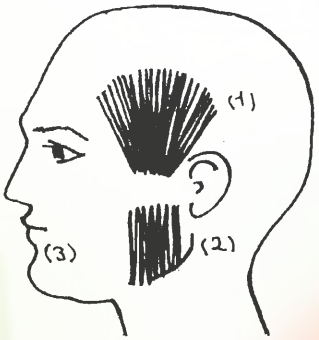
b) Sensory Part:

1. Test for **sensations** including pain (using a pin) and touch (using a piece of cotton) **over the face** and compare between:
 a) Both sides of the face.
 B) The ophthalmic, maxillary and mandibular division on each side.
 c) The inner and outer parts of the face.
2. **Corneal & Conjunctival reflexes** (affer.: ophth. division, Cr 5; effer. Cr 7 bilaterally):
 Ask the patient to look upwards and inwards. Touch the corneo–conjunctival junction from the lateral side (to avoid direct photic stimulation) using a thin piece of cotton.
 * Normally stimulation of one eye results in blinking of both eyes.
 * Absence of blinking on one side denotes facial paralysis of that side.
 * Absence of blinking on both sides denotes:
 - Sensory trigeminal affection of the stimulated side.
 - Bilateral facial paralysis.
 - Organic type of coma.

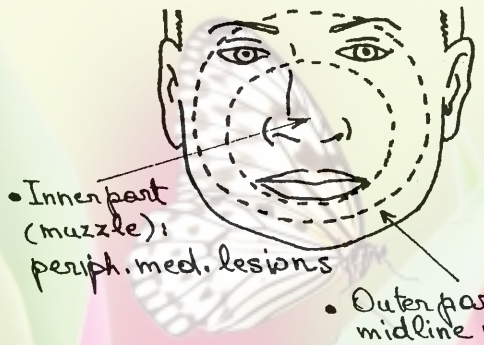
EXAMINATION OF TRIGEMINAL NERVE

I Motor:

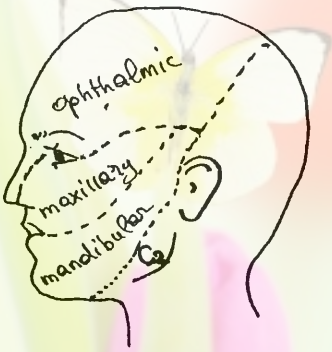
(1) Temporalis } with the jaws clenched
(2) Masseter }
(3) Pterygoids : with open mouth & fixed head.



II Sensory:



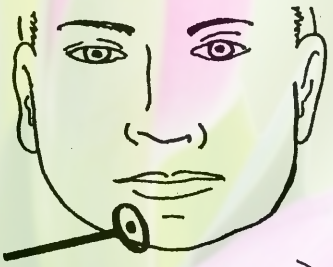
Compare:
1. Both sides
2. Each division
3. Inner & outer parts.



III Reflex:



Corneo-conjunctival (aff. 5 eff 7 bilat.)



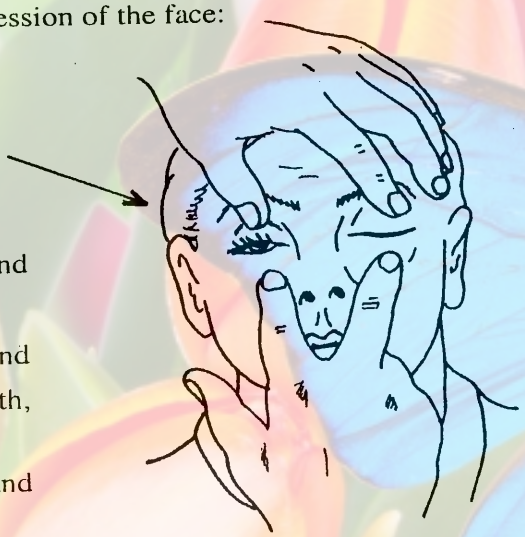
Jaw (aff 5 eff 5)

7. FACIAL NERVE:

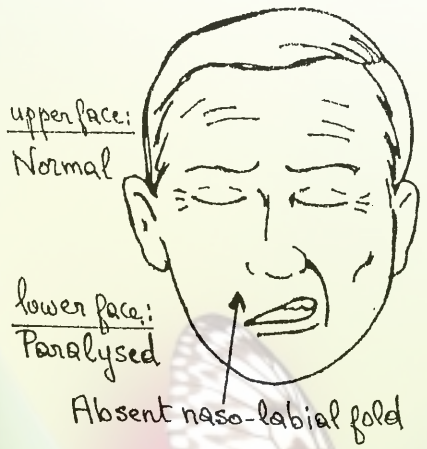
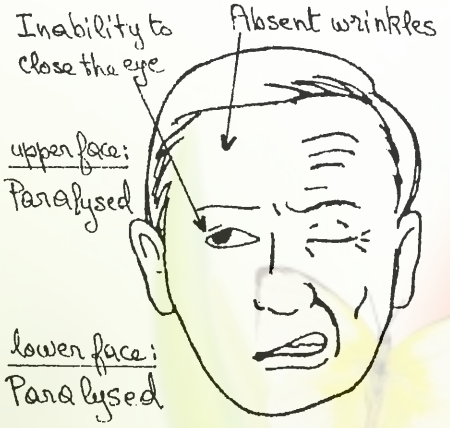
a) Motor Part: Examine for the muscles of expression of the face:

1. Upper face (Frontalis and orbicularis oculi):
 - Test for raising of the eyebrows.
 - Test for firm closure of the eye lids,

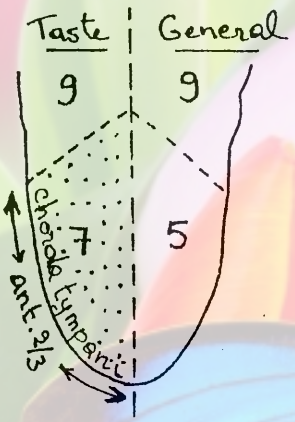
 Bell's phenomena may be seen
2. Lower face (orbicularis oris, buccinator and retractor anguli):
 - Look for absent nasolabial fold and dropping of the angle of the mouth, present in facial paralysis.
 - Test for whistling, blowing the cheeks and showing the teeth.



Differentiation between U.M.N. and L.M.N. facial paralysis

U.M.N.L.	L.M.N.L.
	
1. Paralysis of the muscles of lower half of the face on the opposite side of the lesion.	Paralysis of the muscles of the upper and lower halves of the face of the same side of the lesion.
2. Paralysis involves the voluntary movement but spares the emotional and associative movements.	Paralysis affects voluntary, emotional and associative movements.
3. Paralysis is associated with hypertonia and hyperreflexia.	Paralysis is associated with hypotonia and hyporeflexia.
4. There is associated hemiplegia on the same side of the paralysis.	If there is hemiplegia, it is on the opposite side of the paralysis (crossed hemiplegia).

b) Sensory Part: (Chorda tympani)



Examine for the taste sensation over the anterior two–thirds of the tongue by drying the patient's tongue and then applying a drop of sweet, bitter or salty solution on its tip. See if the patient can properly recognise the taste.

C) Glabellar Reflex:

In the normal adult, tapping the glabella (root of the nose) results in blinking (contraction of orbicularis oculi muscles); this blinking stops after 2–3 taps (due to habituation). In Parkinsonism the blinking continues with the taps as long as the stimulus is applied.

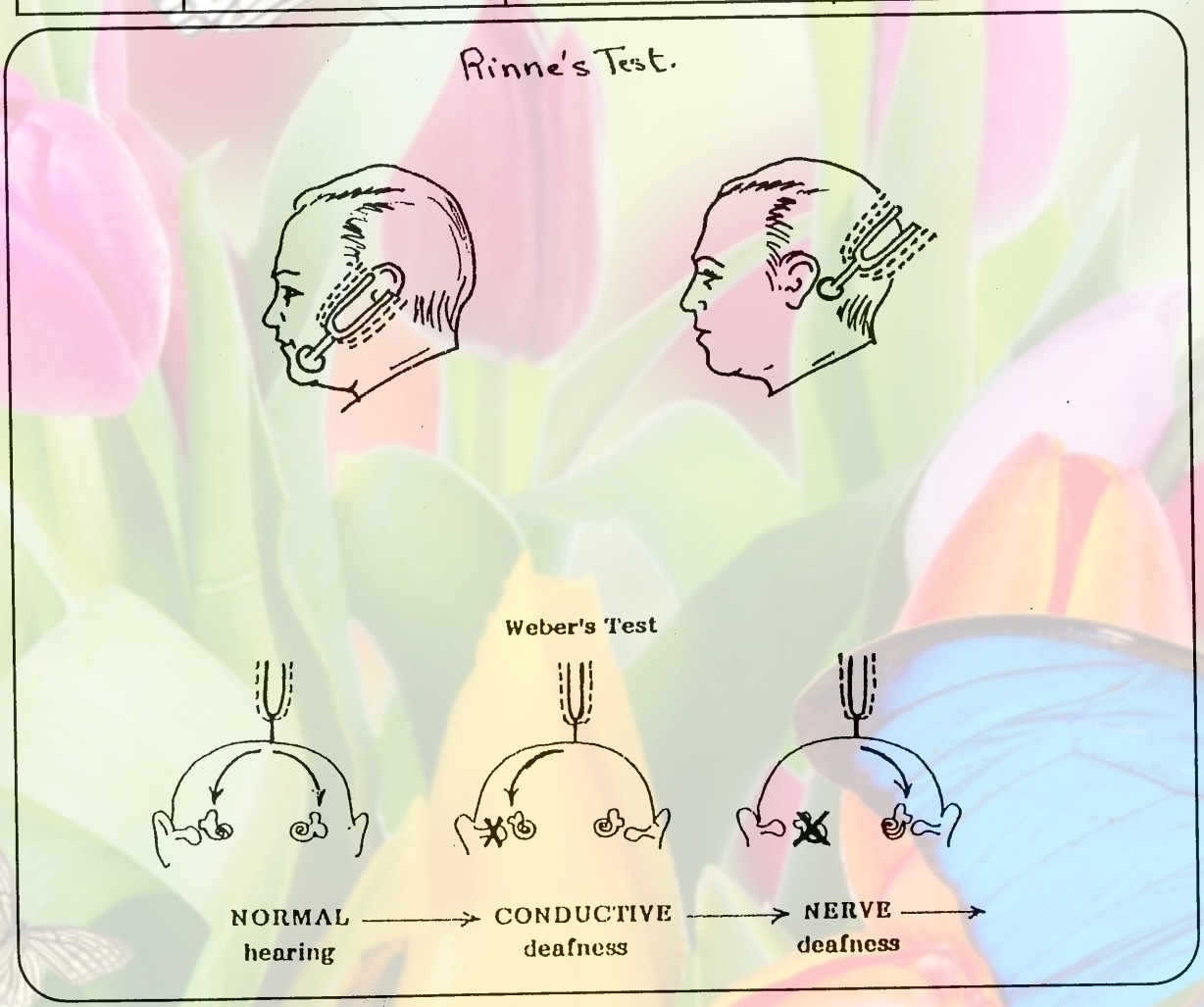


8. COCHLEO-VESTIBULAR NERVE:

a) Cochlear Part: Test for the acuity of hearing using:

1. The Watch test: If there is diminution of the patient's hearing do the following:
2. **Rinne's test**: Using the vibrating tuning fork, compare air conduction (fork placed in front of patient's ear) with bone conduction (fork placed on patient's mastoid process).
3. **Weber's test**: Place the tuning fork in the middle of the forehead.

	Watch test	Rinne's test	Weber's test
Normal	The acuity of the patient's hearing is similar to that of the examiner.	Air conduction is better than bone conduction.	The vibrations are heard in the middle of the forehead.
Nerve deafness	The patient's hearing is less than that of the examiner's.	Both air and bone conductions are diminished.	The vibrations are heard in the normal ear.
Conductive deafness	The patient's hearing is less than that of the examiner's.	Bone conduction is better than air conduction.	The vibrations are heard in the affected ear.



b) Vestibular Part:

Caloric test, rotating chair tests and electronystagmography (E.N.G.).

9. GLOSSOPHARYNGEAL & 10. VAGUS NERVES:



1. Open the patient's mouth and inspect the **uvula**.
 Normally it is central.
 If it is deviated, it is towards the **healthy side**.
2. **Palatal Reflex** (afferent Cr 5, efferent Cr 10).
 Normally stimulation of the soft palate leads to its elevation.
3. **Pharyngeal Reflex** (afferent Cr 9, efferent Cr 10):
 Use 2 tongue depressors, one to depress the tongue & the other to stimulate the pharynx, this results in local contraction & gag reflex.



N.B.:

- Palatal and pharyngeal reflexes are lost in true bulbar palsy and exaggerated in pseudobulbar palsy.
- Isolated lesions of the glossopharyngeal nerve are unknown; it is usually damaged with the vagus and accessory nerves at the jugular foramen.
- The only specific test for isolated vagal lesion is indirect laryngoscopy to observe the position and movements of the vocal cords.

11. ACCESSORY NERVE:

The spinal part is tested by:

- Asking the patient to raise his shoulders against resistance (trapezius).
- Asking the patient to turn his chin against resistance (sternocleidomastoid).



Exam. of trapezius



Exam. of sterno-cleidomastoid

12. HYPOGLOSSAL NERVE:

a. **Inspect** the tongue for:
 1. Deviation: when it is deviated, it is towards the diseased side.
 2. Wasting which indicates a L.M.N.L.
 3. Fasciculations indicate a nuclear lesion as in M.N.D. & syringobulbia. The tongue should be inspected for fasciculations while inside the mouth.



4. Abnormal movements as in Chorea.
5. Dimpling of the tongue on tapping it, in cases of myotonia.
6. Inspection may also reveal:
 - Glazed tongue as in deficiency diseases.
 - Fissured tongue as in mongolism.
 - Ulcers as in Behcet's disease, Herpes simplex.

b. **Test for the power of the muscles of the tongue** by asking the patient to push the inner side of his cheek with the tip of his tongue.

4. EXAMINATION OF THE MOTOR SYSTEM

1. INSPECTION:

a. The **state of the muscles** whether normal, wasted or hypertrophied.

- If there is **wasting** describe it in details:
 - Which limb is affected.
 - Is it unilateral or bilateral.
 - If bilateral, is it symmetrical or asymmetrical.
 - Is it distal more than proximal or vice versa.
- If there is **hypertrophy** report whether it is associated with:
 - Increased power: true hypertrophy.
 - Decreased power: pseudohypertrophy.

b. **Fasciculations** or **fibrillations** (indicating an irritative A.H.C. lesion).

Fasciculation: is a spontaneous contraction of a group of muscle fibres.
It is visible and even palpable.

Fibrillation: is a spontaneous contraction of a single muscle fibre
It is hardly visible except in the tongue.

	Physiological fasciculation	Pathological fasciculation
1. Cause	Anxiety, fatigue, coffee, smoking.	Irritation of AHCs.
2. Type	Coarse.	Fine.
3. Wasting	Absent.	Present.
4. EMG	Normal.	Giant potentials.

c. **Involuntary movements** as chorea, athetosis or tremors; if present, describe them in details i.e., are they static or kinetic, rhythmic or dysrhythmic & what increases or decreases them.

d. **Skeletal deformities** (as pes cavus, hallux valgus, hallux varum . . .) and abnormal positions (as claw hand, drop foot . . .).

e. **Trophic changes** (as loss of hair, brittle nails and ulcers).

2. EXAMINATION OF THE MUSCLE TONE:

Methods of Examination:

a. **Passive** flexion & extension of all the joints.

b. **Shaking** method for the wrist & ankle only.

c. **Gower's method** for the shoulder (as in myopathy): Place your hands in the patient's axillae & try to lift his shoulders.

Abnormal Muscle Tone may be:

a. Decreased: **Hypotonia (Flaccidity)** which may be due to:

1. Lower motor neurone lesions.
2. Shock stage of acute U.M.N.L.
3. Rheumatic chorea.
4. Cerebellar lesions.
5. Posterior column lesions.
6. Hypotonic form of cerebral palsy.

b. Increased: **Hypertonia** which may be due to:

1. Pyramidal (U.M.N.) lesion = Spasticity; the hypertonia is:
 - "**Clasp–knife**" where the initial resistance to movement is suddenly overcome.
2. Extrapyramidal lesions other than chorea = Rigidity; the hypertonia may be:
 - "**Lead pipe**" where there is a steady increase in resistance, or
 - "**Cog–wheel**" where the resistance is intermittent.

	Spasticity	Rigidity
Site of lesion	Pyramidal	Extrapyramidal
Distribution	– Distal >proximal – Flexors of U.L. – Extensors of L.L. & trunk	Proximal >distal Flexors of U.L., L.L. & trunk
Character	Clasp–knife	Lead pipe or cog–wheel
Deep reflexes	Hyperreflexia	Hyporeflexia

N.B.: Other causes of Hypertonia

3. Myotonia.
4. Catatonia.
5. Meningeal irritation.
6. Hysterical.

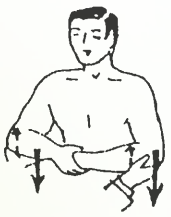
3. EXAMINATION OF THE MUSCLE POWER:

The muscles are tested against resistance on condition that they are not totally paralysed i.e. the patient can actively contract them.

a) IN THE UPPER LIMB:

Shoulder (mainly C4–C5):

* **Adduction**: pectoralis major & minor assisted by latissimus dorsi & teres major.
 Ask patient to adduct his arms against resistance; or while patient presses his hands to his waist, palpate the anterior axillar fold for the contracted pectoralis.



Exam. of shoulder adductors

* **Abduction**

0°–150° suprasinatus
15°–90° deltoid
90°–180° trapezius
} Ask the patients to lift His arm straight out at Right angles to his side.



Exam. of shoulder abductors

* **Flexion**: anterior fibres of deltoid; ask patient to raise his arm forwards against resistance.
* **Extension**: posterior fibres of deltoid; ask patient to raise his arm backwards against resistance.
* **Lateral rotators**: infraspinatus & teres minor.
* **Medial rotators**: latissimus dorsi & subscapularis.
* **The serratus anterior**: ask patient to push his arm forwards against resistance; paralysis of this muscle leads to winging of the scapula.



Exam. of elbow flexor "biceps"



Exam. of elbow flexor "brachioradialis"

Elbow (C 5, 6, 7):

* **Flexors**: biceps, brachialis & brachioradialis.
 – Biceps: with the patient's arm extended by his side & the hand fully supinated, ask him to flex his elbow against resistance.
 – Brachioradialis: as for the biceps but with the hand semi–pronated.



Exam. of elbow extensor "triceps"

* **Extensors**: triceps: with the patient's elbow flexed ask him to extend it against resistance.

Wrist (C 7, 8): Test for flexion and extension against resistance while the fist is closed.

Hand (C 8, Th 1):

- *Thumb:*
 1. Opponens pollicis: ask the patient to touch the tip of his little finger with the tip of his thumb.
 2. Abductor pollicis brevis: it is the only muscle of he hand supplied by the median nerve than can be easily tested (as in carpal tunnel syndrome). Ask the patient to abduct his thumb at a right angle to the palm of the hand; the muscle can be seen and palpated.

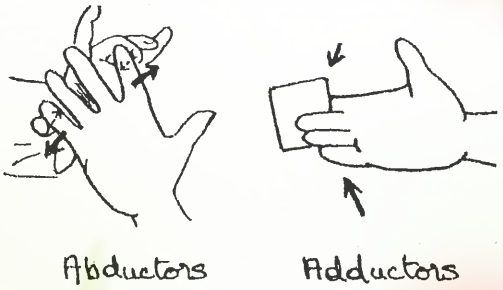


Exam. of Opponens pollicis



Exam. of Abductor pollicis brev.

- *Other fingers:*
 1. Abductors: Dorsal interossei; patient abducts his fingers against resistance.
 2. Adductors: Palmar interossei; patient grabs a paper between 2 fingers.
 3. Lumbricals: Patient puts his fingers in the writing position.



b) ABDOMINAL MUSCLES: (Th6–Th12)L are tested for specially in cases of myopathy. The patient lies down, puts his hands over his chest, then attempts to sit up.

c) IN THE LOWER LIMB:

Hip:

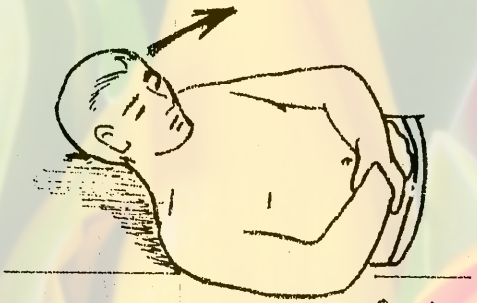
* **Flexion**: ileo–psoas (L1, 2): ask the patient to flex his hip against resistance.
* **Extension**: gluteus maximus (L5, S1,2): with the patient lying face downwards in bed, fix his trunk with your hands & ask him to raise his L.L. against resistance.
* **Adduction**: adductors longus, brevis & magnus (L2,3,4) assisted by pectineus & gracilis. Abduct the thigh & ask the patient to bring it towards the midline.
* **Abduction**: gluteus medius & minimus (L5, S1) while the thigh is in the midline, ask the patient to move it outwards.

Knee:

* **Extension**: quadriceps (L2, 3, 4): ask the patient to maintain knee extension while you try to bend the knee; or bend the patient's knee & ask him to straighten it.
* **Flexion**: hamstrings (S1, 2): ask the patient to pull his heel towards his buttock against resistance.

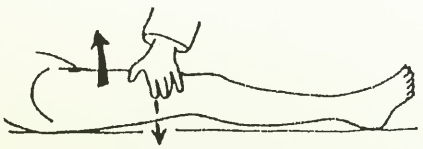
Ankle:

* **Dorsiflexion**: ant. tibial group (L4, 5).
* **Plantar flexion**: calf muscles (S1, 2): the patient moves his foot upwards & downwards against resistance.
* **Inversion**: tibialis anterior & posterior (L4).
* **Eversion**: peroneal muscles (L5): the patient inverts & everts his foot against resistance.

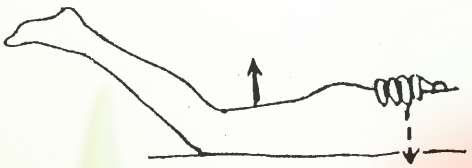

Exam. of Abdominal Muscles.

EXAMINATION OF THE MUSCLES OF L.L.

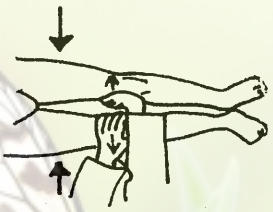
I Hip:



Flexion



Extension

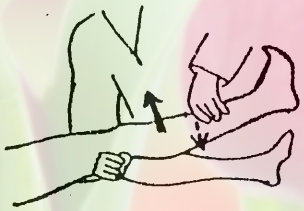


Adduction



Abduction

II Knee:



Extension



Flexion

III Ankle:



Dorsiflexion



Plantarflexion



Inversion



Eversion

4. EXAMINATION OF THE REFLEXES:

a) DEEP REFLEXES:

In the Upper Limb:

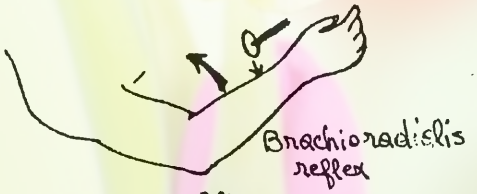
1. **Biceps reflex** (C 5, 6):

 Elicited by a tap upon the biceps tendon while the elbow is at 120°. The tap is done on your index finger placed over the tendon. It results in mild contraction of the biceps with slight flexion of the elbow.



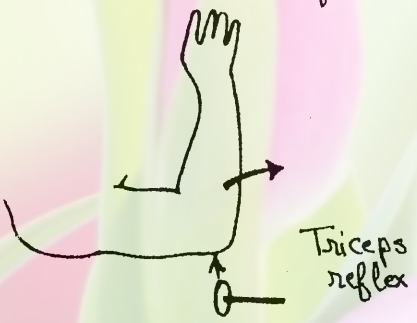
2. **Brachioradialis reflex** (C 5, 6):

 Elicited by a tap 3–4 cm above the styloid process of the radius, while the elbow is at 120°. It results in mild contraction of the brachioradialis and slight flexion of the elbow.



3. **Triceps reflex** (C 6, 7):

 Elicited by a tap directly on the triceps tendon while the elbow is flexed at 90°. It results in mild contraction of the triceps with slight extension of the elbow.



4. **Supraspinatus** (C 3, 4) and **finger** (C8, Tl) **reflexes**:

 They are normally absent. If present, they indicate an U.M.N.L.

 * The **supraspinatus** reflex is done by tapping the supraspinatus muscle; in U.M.N.L. there is visible contraction of the muscle with slight abduction of the shoulder.
 * The **finger** reflex is done by tapping the palmar surface of the middle 3 fingers while they are slightly flexed; in U.M.N.L. there is prompt flexion of the fingers.

In the lower limb:

1. **Knee reflex** (L 2, 3, 4):

 Elicited by a tap on the quadriceps tendon while the hip joint is slightly flexed and the knee joint is flexed and supported from beneath by your hand. It results in visible contraction of the quadriceps and extension of the knee.



2. **Ankle reflex** (S 1, 2):

Elicited by a tap on the tendon Achilles while the thigh is abducted and externally rotated, the knee is flexed at 90° and the ankle is dorsiflexed by the examiner. It results in mild contraction of the calf muscles with plantar flexion of the ankle.



3. **Patellar** (L 2, 3, 4) and **Adductor** (L 4) **reflexes**:

They are normally absent If present, they indicate an U.M.N.L.

* The **patellar** reflex is done by pressing the upper border of the patella downwards with the examiner's index finger and then tapping the finger with the hammer; in U.M.N.L. there is contraction of the quadriceps and upward displacement of the patella.
* The **adductor** reflex is done by tapping the index finger placed just above the adductor tubercle, while the hip is externally rotated and slightly abducted; in U.M.N.L. there is visible contraction of the adductors with adduction of the thigh.

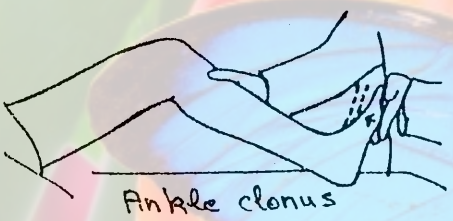
N.B.:

* While eliciting the deep reflexes one should observe the movement of the joint as well as that of the acting muscles which, therefore, should be bared.
* Don't report that a deep reflex is absent unless the patient does a **reinforcement** by clenching his teeth or clutching his hands together (Jendrassik's manoeuvre).
* If there is hyperreflexia, try to elicit clonus.

CLONUS: is a rhythmical series of contractions in response to the sudden sustained stretch of the tendon of the muscle. Clonus may be:

1. **Organic** denoting a definite U.M.N.L. in which case it stops with release of the stretch of the muscle.
2. **Hysterical** where it persists in spite of release of the stretch of the muscle.

* **Ankle clonus** is obtained by passive plantar flexion of the joint followed by sudden dorsiflexion.
* **Patellar clonus** is obtained by holding the patella and displacing it slightly upwards; this is followed by a sudden downward displacement of the patella.
* **Wrist clonus** is obtained by sudden and sustained extension of the wrist.





b) SUPERFICIAL REFLEXES:

They are absent in:

- U.M.N.L. above the level of the segmental supply of the reflex.
- L.M.N.L. affecting the reflex arc itself.

1) Abdominal Reflexes (Th6 – Th12):

- Upper abdominal reflex (Th6–10): light stroking of the skin of the abdomen above the umbilicus, from the periphery inwards, using a pin.
- Lower abdominal reflex (Th10–12): light stroking is done below the level of the umbilicus, also from the periphery inwards.

In both cases, contraction of the ipsilateral abdominal muscles can be seen.



2) Cremasteric Reflex (L1): elicited by a stroke with a pin, along the upper part of the medial aspect of the thigh resulting in visible contraction of the cremasteric muscle.

3) Gluteal Reflex (L4, 5): elicited by stroking across one of the buttocks with a pin resulting in contraction of the ipsilateral gluteal muscles.

4) Anal Reflex (S3, 4, 5): elicited by scratching the skin of the perineal region resulting in contraction of the external anal sphincter.

5) Plantar Reflex (S1, 2)

Normally, stroking the sole of the foot with a blunt object results in plantar flexion of the toes. If there is dorsiflexion, with or without fanning of the toes, it denotes an U.M.N.L. However, dorsiflexion may occur physiologically in deep sleep and in infants below one year. It can be elicited by the following methods:

1. **Babinski method**: A scratch is made on the lateral aspect of the sole of the foot from the heel towards the toes.
2. **Shaddock's method**: A scratch is made on the lateral aspect of the dorsum of the foot from the lateral malleolus to the little toe.
3. **Oppenheim's method**: Firm pressure is applied on the skin over the lower part of the shaft of the tibia, from above downwards.
4. **Gordon's method**: The calf muscles are firmly squeezed.
5. **Schaefer's method**: The tendon Achilles is firmly squeezed.
6. **Gonda's method**: The 3rd & 4th toes are passively flexed, then suddenly released.



5. EXAMINATION OF THE SENSORY SYSTEM

A. SUPERFICIAL SENSATIONS including pain, temperature & touch:

We examine for pain using a pin & for touch using a piece of cotton.

- Compare both sides leg to leg, arm to arm & face to face.
- Compare on each side, the L.L. with the trunk, with the U.L. & with the face.
- In case of hyposthesia in a limb, test all around it to differentiate between radicular sensory loss & glove & stock hyposthesia.

SENSORY SUPPLY OF THE BODY:

C2	Angle of jaw, lateral aspect of neck
C3, 4	Shoulder, down to manubrium
C5	Lateral aspect of arm
C6	Lateral aspect of forearm, thenar eminence & thumb
C7	Middle aspect of forearm, middle of the palm, middle 3 fingers
C8	Medial aspect of forearm, hypothenar eminence & little finger
T1	Medial aspect of arm
T2–T7	Thorax (T4 → nipple)
T8–T12	Abdomen (T10 → umbilicus) (T12 → inguinal ligament)
L1	Upper $^1/_3$ front of thigh
L2	Middle $^1/_3$ front of thigh
L3	Lower $^1/_3$ front thigh
L4	Anterolateral aspect of thigh, front of knee, anteromed. aspect of leg, med. aspect of foot & big toe
L5	Lat. aspect of thigh, lat. aspect of leg, middle $^1/_3$ of dorsum of foot & middle 3 toes
S1	Posterolateral aspect of thigh & leg, lateral $^1/_3$ of dorsum of foot & little toe
S2	Posterior aspect of thigh, leg & sole of foot
S3, 4, 5	Anal, perianal & gluteal region (saddle shaped area) in concentric manner

B. DEEP SENSATIONS: they include:

1. **Vibration sense**: place the vibrating fork over the bony prominences: medial malleolus – ant. tibial tubercle – ant. superior iliac spine (A.S.I.S.) – clavicle.
 Ask the patient if he feels the fork's vibrations & if they are felt equally on all sites
 If V.S. is diminished or lost over med. malleolus, check A.S.I.S.; if lost, it suggests posterior column lesion; if intact, it suggests P.N. lesion
2. **Joint sense** (sense of position and movement): First show the patient, with his eyes open, the position of his big toe (dorsi–flexed, plantar–flexed); then with his eyes closed, move the big toe and ask him if he feels it moving and if so in which direction. The big toe should be caught gently, from the sides.
3. **Muscle sense**: by pinching the calf. The muscle sense may be normal where the patient feels a disagreeable sensation. It may be lost (Abadie's sign) as in neurosyphilis or exaggerated (tender calf). (See page 81).
4. **Nerve sense**: by pressing the ulnar nerve and the lateral popliteal nerve against the bones. Normally, it results in an electric–like sensation.
5. **Romberg's test**: ask the patient to stand with the heels together, 1st with his eyes open, then with his eyes closed.
 Note any swaying or loss of balance. If present:
 - With eyes open or closed = cerebellar ataxia.
 - Only with closed eyes = sensory ataxia.

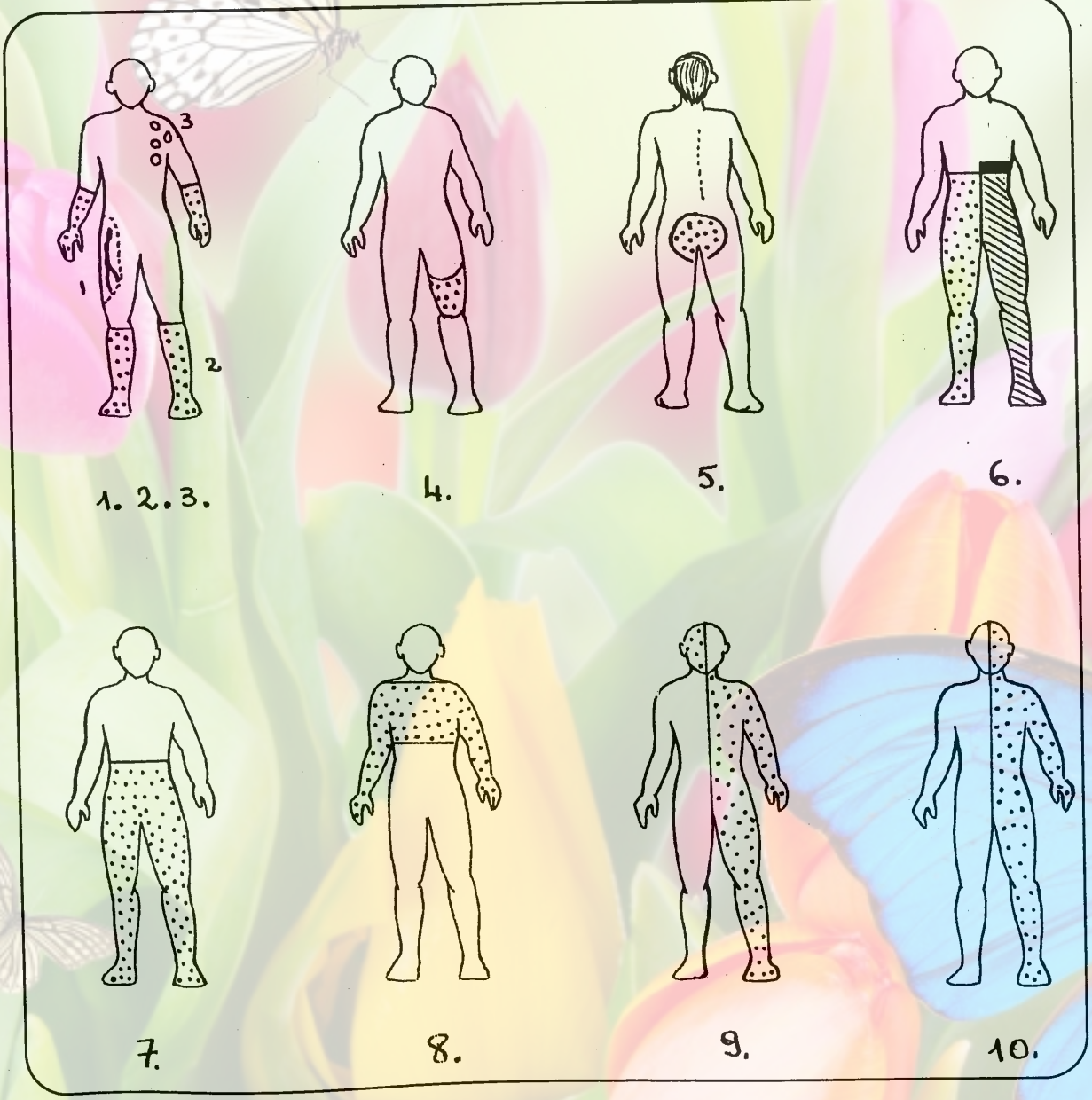
C. CORTICAL SENSATIONS:

They are only examined when the superf. & deep sensations are intact.

1. **Tactile localisation**: ask the patient to close his eyes; then prick his finger & ask him to localise the site of the prick.
2. **Two–points discrimination**: with the patient's eyes closed, deliver 2 simultaneous pricks, e.g. on the finger (5 mm apart) or on the legs (4 cm apart). Normally the 2 pricks are felt distinct from each other.
3. **Stereognosis**: with his eyes closed, the patient is asked to recognise a familiar object placed in his hand.
4. **Graphosthesia**: with his eyes closed, the patient is asked to recognise a number or letter drawn over his palm.
5. **Perceptual rivalry**: normally if you deliver 2 simultaneous pin pricks at 2 corresponding sites of the body, both pricks are felt; in cortical sensory loss, only the prick on the healthy side is felt.

PATTERNS OF SENSORY LOSS

Pattern of sensory loss	Site of lesion
1. Mononeural 2. Stock & glove 3. Maculo–anaesthetic (leprosy)	Peripheral nerve
4. Radicular sensory loss	Root
5. Saddle area loss	Conus
6. Dissociated sensory loss (Brown–Sequard syndrome)	Unilateral cord lesion
7. Sensory level	Extramedullary lesion
8. Jacket sensory loss (dissociated)	Intramedullary lesion
9. Crossed hemihyposthesia	Lateral medullary syndrome
10. Hemihyposthesia	Capsular & brain stem lesions
11. Cortical sensory loss	Area (1, 2, 3) of parietal lobe

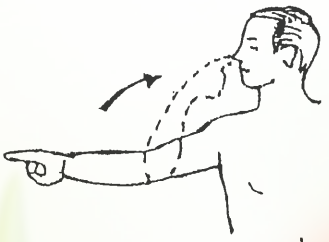


6. EXAMINATION OF COORDINATION

A. IN THE UPPER LIMB:

1. **Finger–to–nose test**: The patient brings the tip of his forefinger from a distance onto the tip test is conducted with the eyes open the
2. **Finger–to–finger test**: The patient brii forefinger from the distance of his out meet each other in the midline.
3. **Finger–to–doctor's finger test**: The p tip of his forefinger from a distance o doctor's forefinger.

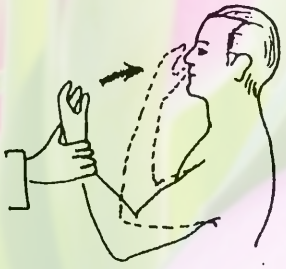
 In any of the above tests you may find:
 a) Decomposition of movement.
 b) Kinetic intention tremors whicl evident as the patient's forefinge target.
 c) Dysmetria in the form of Hypometria.
4. **Adiadokokinesis or Dysdiadokokines** asked to do rapidly alternating moveme and supination of the forearm. In cereb is failure to perform the movements.
5. **Rebound phenomenon**: The patient, fixed, flexes it against resistance. Whe suddenly released the patient's forearm may hit his face or shoulder.
6. **Buttoning and unbuttoning test**: earli



Finger-to-nose test



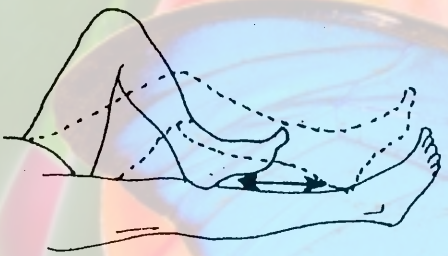
Adiadokokinesis



Rebound phenomenon

B. IN THE LOWR LIMB:

1. **Heel–to–knee test**: The patient raise down its heel onto the knee of his othe down along the shaft of the tibia.
2. **Walking** along a straight line, foot unilateral cerebellar lesions, there is diseased side.
3. **Romberg test**: Ask the patient to sta together. Swaying or loss of balance eyes are open or closed.



Heel-to-Knee test.

7. EXAMINATION OF THE BACK AND SPINE:

Examine for:

1. Tenderness.
2. Deformity.
3. Hair tuft.
4. Swelling.
5. Abnormal pigmentation.

8. EXAMINATION OF THE CRANIUM:

Examine for:

1. Size, shape, sutures & fontanelles.
2. Bony bosses & tenderness.
3. Dilated veins, bruits & naevi.
4. McEwen's sign in brain tumours.

9. EXAMINATION OF THE NECK:

Examine for:

1. Signs of meningeal irritation: neck retraction – +ve Brudzinski neck sign.
2. Bruit over the neck as in carotid artery stenosis.

10. EXAMINATION OF THE GAIT:

If the patient can walk:

Lesion	Cause	Gait
1. U.M.N.L. (Δ) a. Unilateral b. Bilateral	Hemiplegia Paraplegia	Circumduction Scissor
2. L.M.N.L a. Periph. Nerve b. Muscle	P.N. Myopathy	High steppage Waddling
3. Post. column	S.C.D. & tabes dorsalis	Stamping "Strike forcibly the ground"
4. Cerebellum a. Archicerebellum b. Neocerebellum • Unilateral • Bilateral	Friedriech's ataxia Cerebellar astrocytoma Marie's ataxia	Wide base "drunken" Deviation to one side Zigzag
Extra Δ	Parkinsonism: Mild Severe Chorea	Short steppage Shuffling or festinant Dancing
Hysterical	Neurosis	Astasia Abasia